DISEASES OF THE SPINAL CORD

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To

THE PAST AND PRESENT STUDENTS

OF

THE MANCHESTER MEDICAL SCHOOL

PREFACE

The following account of diseases of the spinal cord is based on notes of lectures, which I have given at the Manchester Medical School during the last fifteen years. To these notes numerous additions have been made, and the lecture form has been altered.

The work is not intended to furnish an exhaustive account of spinal diseases; but it is hoped that it may be of service as a *text-book* and as an introduction to the study of the subject.

The illustrations are (with one exception) from my own drawings and photographs, or from micro-photographs of my own sections. For Fig. 174 I am indebted to the kindness of Professor Homén, of Helsingfors.

Though I have tried to fairly acknowledge the work of previous writers, space limits only allow reference to the sources of a small proportion of the statements made; and, therefore, it is necessary for me to mention how greatly I am indebted to the earlier works of the late Dr. James Ross, of Manchester, and to the text-books and writings of Sir Wm. Gowers and Dr. J. Taylor, and to the works of Professors Oppenheim, Strümpell, Schultze, Leyden and Goldscheider, Dejerine and Thomas, Marie, and Allen Starr.

In the pathological anatomy of spinal diseases I am specially indebted to the work of Schmaus and Sacki, *Vorlesungen über die pathologische Anatomie des Rückenmarks*, Wiesbaden, 1901, and to the *Handbuch der pathologischen Anatomie des Nervensystems*, edited by Flatau, Jacobsohn and Minor, Berlin, 1903.

In the preparation of this work a bibliography of spinal diseases was commenced; but it was soon evident that, owing to the very extensive literature of the subject, the size of the book would have been very greatly increased thereby. For this and other reasons, I decided to restrict the references to the articles and works which I have found of most service. Senior students, or readers who may not be well acquainted with the literature of spinal disease, will perhaps find these references useful. To such readers the text-books already mentioned

may be recommended; and for an account of the most recent work the abstracts and articles in the following journals may be consulted: *The Review of Neurology and Psychiatry* (Edinburgh); the *Neurologisches Centralblatt* (Leipzig); *Brain* (London); *The Medical Chronicle* (Manchester).

Amongst the references I have frequently included my own abstracts and reviews which have appeared in the *Medical Chronicle*; not because I considered that they could have any claim to be regarded as important articles on the subjects, but simply because they have contained abstracts of, and numerous references to, the recent neurological work of others. These reviews may be of service to those who are working up the literature of any section.

Some knowledge of the more important methods of staining of microscopic sections of the cord is necessary in order to understand many of the illustrations, and at the request of the publishers I have added, as an appendix, a short account of a number of the simpler processes. This account may be useful to readers who are commencing to work at the pathological histology of diseases of the spinal cord. But for a description of many of the numerous recent complicated methods the reader may be referred to the text-books mentioned in the appendix.

For valuable assistance in correcting the proofs I am indebted to Dr. J. D'Ewart.

R. T. W.

MANCHESTER,
1908.

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DISEASES OF THE SPINAL CORD

SECTION I

STRUCTURE OF THE SPINAL CORD

In order to understand the symptoms and localisation of spinal diseases, a knowledge of the more important facts of the anatomy and physiology of the spinal cord is necessary. The following outline of the structure and functions of the spinal cord will refer, however, to those points only which are of practical importance in the study of spinal diseases.

The spinal cord commences just below the medulla oblongata, at the origin of the first cervical nerves, and extends, within the vertebral canal, down to the level of the upper border of the second lumbar vertebra. Below this level the nerve roots of the cauda equina lie within the vertebral canal (Fig. 86A and 86B, pp. 182–183).

The cord is closely invested by an adherent connective tissue sheath, the pia mater. External to this, are the delicate sheath, the arachnoid, and the thick outermost sheath, the dura mater. The space between the dura mater and pia mater is occupied by cerebro-spinal fluid.

The dura mater is separated from the bony wall of the vertebral canal by adipose tissue and plexuses of veins. The dura mater extends along the nerve roots for a short distance, in the form of a sheath.

The external form and enlargements of the cord are described in textbooks of anatomy and do not require consideration here.

The portion of the spinal cord from which each pair of nerves arises is spoken of as a segment. (To these spinal segments the term of metameres is also applied.)

General Histology.

A transverse section of the cord shows the white substance surrounding the central grey matter. The anterior median fissure and the posterior median septum divide the white matter into two lateral portions (*see* Fig. 1). The relation and shape of white and grey matter vary at different levels; but according to A. Bruce there is a type of outline of the anterior horn which is characteristic of each spinal segment in the cervical and lumbar enlargements, and the investigations of E. Bramwell confirm this view.[1]

[1] For illustrations, *see* E. Bramwell's article, *Review of Neurology and Psychiatry*, May, 1906.

The *white substance* is composed of medullated nerve fibres, supported and embedded in a form of connective tissue—the neuroglia. A large portion of these nerve fibres run in a longitudinal direction. Hence a

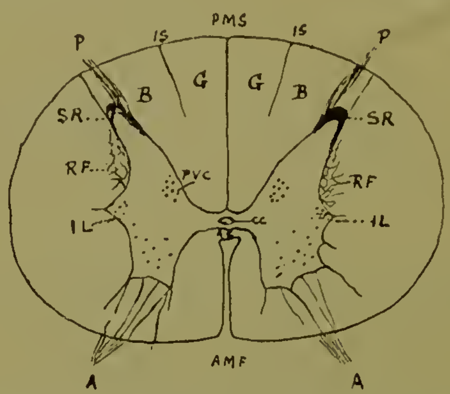


Fig. 1.—Transverse Section of Cord (Dorsal Region).

PMS = Posterior median septum.
AMF = Anterior median fissure.
cc = Central canal.
ac = Anterior commissure.
A = Anterior root.
P = Posterior root.
IS = Intermediate septum.
G = Column of Goll (between IS and PMS).
B = Column of Burdach (between IS and posterior root and horn).
PVC = Posterior vesicular column of Clarke.
RF = Reticular formation.
IL = Intermedio-lateral grey column.
SR = Gelatinous substance of Rolando.

transverse section of the white matter reveals cross sections of the axis cylinders of the nerve fibres, each surrounded by a myelin sheath, but external to this there is no neurilemma sheath. The supporting neuroglia tissue consists of fibres and nucleated cells. Only a small zone of protoplasm surrounds the nucleus of these neuroglia cells (*see* Fig. 2).

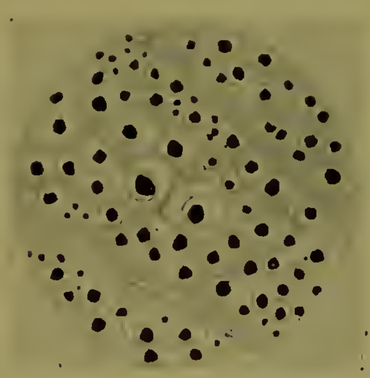


Fig. 2.—White matter of Cord (formol and silver nitrate stain). Nerve fibres in transverse section. Axis-cylinders deep black.

The nerve fibres and surrounding neuroglia lie in a coarser scaffold-work, formed by the branching processes of connective tissue septa which pass inwards from the pia mater. Blood vessels run in the septa and in the neuroglia tissue.

The *grey substance* of the spinal cord consists of (1) a thick felt-work of fine medullated nerve fibres (stained black by the methods of Weigert and Pal); (2) of nerve cells and their processes; (3) of neuroglia tissue; (4) of numerous small blood vessels (*see* Figs. 3A and 3B).

The grey matter of each half of the cord consists of anterior and posterior horns; and to the part between the two the name of intermediate grey substance is given. The posterior horn reaches to the surface of the cord. The grey matter of the two halves of the cord is connected by a grey commissure, in the centre of which is the central canal. In the

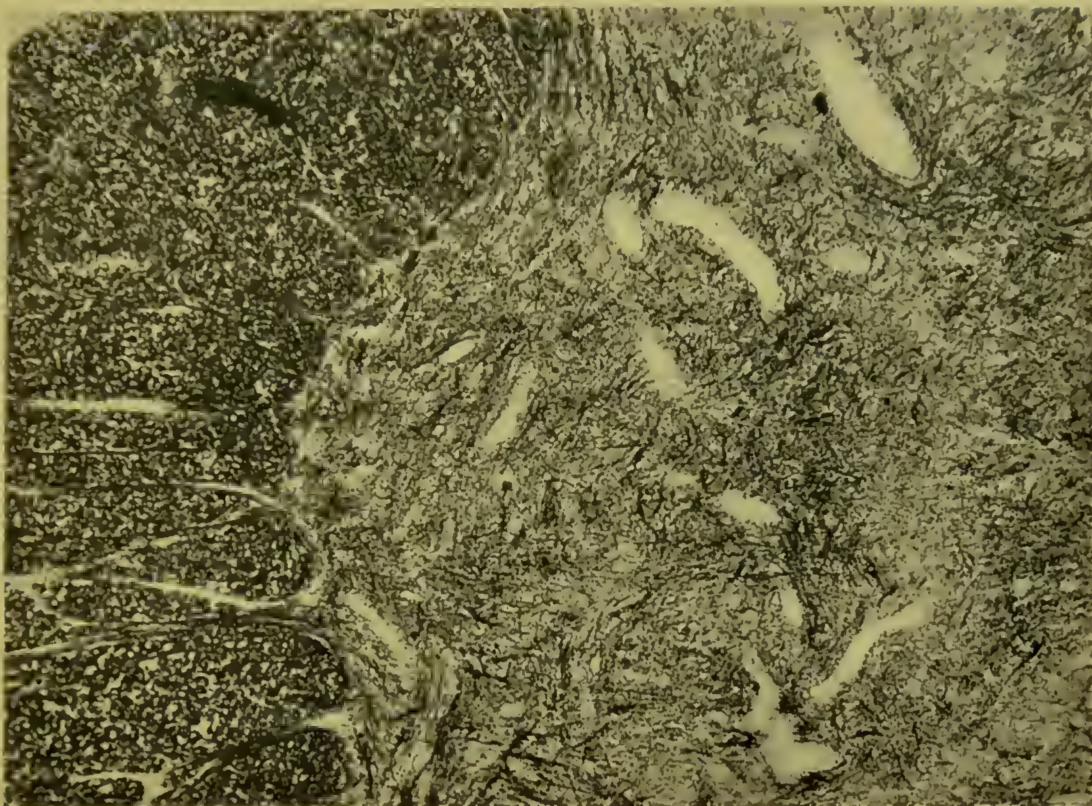


FIG. 3A.—Grey Matter of Anterior Horn of Spinal Cord (Pal's method of staining) showing felt work of fine nerve fibres with myelin sheath, stained blue-black. Around the anterior horn is the white matter in transverse section (to the left of the illustration).

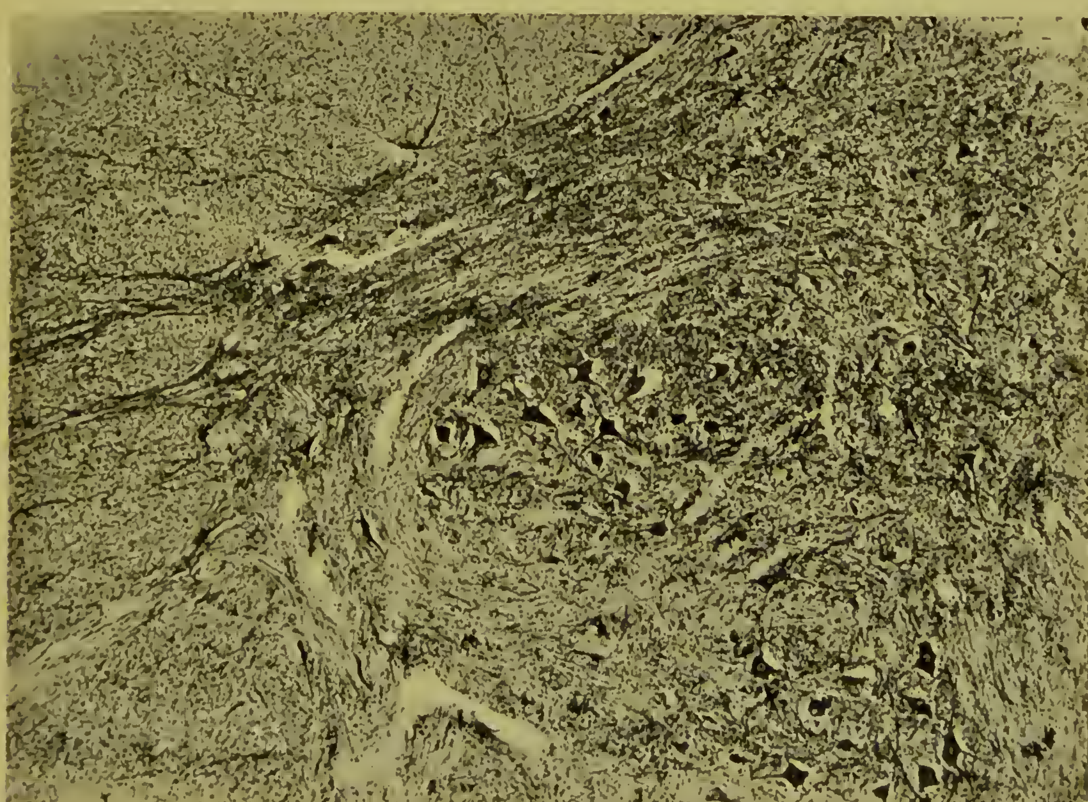


FIG. 3B.—Anterior Horn of Grey Matter of the Spinal Cord (formol and nitrate of silver method) showing nerve cells and axis cylinders of fine nerve fibres stained black.

lower dorsal and upper lumbar regions (from the level of the eighth dorsal to the second lumbar nerves), a group of nerve cells is situated at the inner part of the neck of each posterior horn— the posterior vesicular column of Clarke. Most of the cells are fusiform and placed vertically. Nerve fibres pass from and into this column of cells.

The groups of nerve cells in the anterior horns are described on p. 108, and the grey intermedio-lateral tract on p. 58.

The fibres of the anterior roots proceed from the anterior horns of grey matter. Many fibres of the posterior nerve roots enter the posterior horns as will be described subsequently.

The form and position of the grey substance, the anterior and posterior horns, the substantia gelatinosa of Rolando, the grey and white commissures, the anterior, lateral and posterior columns, the anterior and posterior nerve roots and the central canal are shown in Fig. 1. (For a detailed account of the coarse histology of the cord the reader is referred to text-books of anatomy and histology.)

The methods of histological examination which are of most value for revealing the finer structure of the spinal cord require brief mention. (For a detailed account of the methods *see* Appendix, p. 412, and writer's article "Notes on Methods of Pathological Examination of the Spinal Cord," *Med. Chron.* Feb. 1904.)

The external form of nerve cells and their numerous processes is beautifully demonstrated by Golgi's method and its modifications, in which the cell and its process are stained deep black, by a prolonged immersion of the tissue in a solution of nitrate of silver or corrosive sublimate. But this method is of service chiefly in studying the normal histology, and is often unsatisfactory in pathological work. The formol and nitrate of silver method is useful in pathological work; nerve cells and their processes and the axis cylinders of nerve fibres are stained deep black by this method. In certain modifications of the method, fibrillæ in the axis cylinders and in the nerve cells can be detected (Ramon y Cajal, *Comp. Rend. Soc. Biol.* Dec. 12, 1903).

The internal structure of nerve cells is demonstrated by Nissl's methylene blue method and by its simple modification—the toluidin-blue stain.

For the staining of nerve fibres two methods are of greatest value—those of Weigert and Marchi.

In *Weigert's* method (copper-haematoxylin), and in its simple modification—Pal's method, the outer part of the myelin sheath of *normal* nerve fibres is stained deep blue-black. Neuroglia, blood vessels, and nerve cells are stained yellow or yellowish-brown in Weigert's method, stained red in Pal's method.

In *Marchi's* method (osmic acid and Müller's fluid) normal fibres are not stained, but the degenerated products of *diseased* fibres are coloured deep black. Thus normal fibres appear black in sections prepared according to Weigert's and Pal's methods and tracks of degeneration

are yellowish brown; whilst in sections stained according to Marchi's method the degenerated fibres are black, the other structures yellow.

The formol and nitrate of silver method is useful in demonstrating clearly the axis cylinders, which are coloured deep black, whilst the myelin sheath is not stained. The writer's modification is a simple method of carrying out this process (*see* Appendix). Plate I illustrates these methods of staining.

THE HISTOLOGY OF NERVE CELLS.[1]

The *external form* and the processes of nerve cells can be best studied by the method of Golgi, and the formol and silver nitrate methods. The nerve cells and their processes appear black and opaque in sections stained according to these methods. In the adult all nerve cells (in

FIG. 4.—Three Nerve Cells of the Spinal Cord (diagrammatic). The cell on the left stained deep black according to Golgi's method. *a* = axis-cylinder process or axon; other processes = dendrites *d*. Cell in centre stained with formol and silver nitrate. Fibrillæ in cell and processes demonstrated. Cell to the right stained according to Nissl's method.

man) have processes; nerve cells without processes are only found in the embryo. According to the number of processes, nerve cells are described as unipolar, bipolar, or multipolar.

Unipolar cells are rare; in the human adult they are found in the cerebro-spinal ganglia (ganglia of posterior nerve roots). In the unipolar nerve cells of the posterior ganglia the process of the cell soon divides into a central and peripheral fibre. *Bipolar* cells are found almost exclusively in the peripheral sensory nervous system. *Multipolar* cells are the most numerous. Their processes are of two kinds—protoplasmic processes or dendrites, and axis-cylinder processes or axons. The dendrites have an irregular contour; they divide and subdivide into numerous branches, and gradually become more narrow. The axis-cylinder process is almost always single; it often becomes ultimately the axis-cylinder of a nerve fibre. The axis-cylinder process preserves in its entire course a slender, smooth contour; its calibre is as a rule uniform, and is maintained for a considerable distance from the cell; its course is usually direct, so that

[1] This account of the structure and pathological histology of nerve cells is based on the work of Van Gehuchten, Marinesco, Schmaus, and others, and refers mainly to the nerve cells of the spinal cord.

PLATE I.

METHODS OF STAINING NERVE CELLS AND FIBRES.

I. Nerve cells of the spinal cord. *Toluidin-blue stain.*
(1) Normal cells, showing Nissl's granules.
(2) Cells, showing chromatolysis.
(3) Cells, showing peripheral chromatolysis.

II. Normal white matter of the spinal cord. *Weigert's (or Pal's) stain.* Transverse section.

III. Normal grey matter of spinal cord. *Weigert's (or Pal's) stain.*

IV. Transverse section of spinal cord. *Weigert's stain.* Disseminated sclerosis. A = Normal nerve fibres. B = Patch of sclerosis; nerve fibres absent.

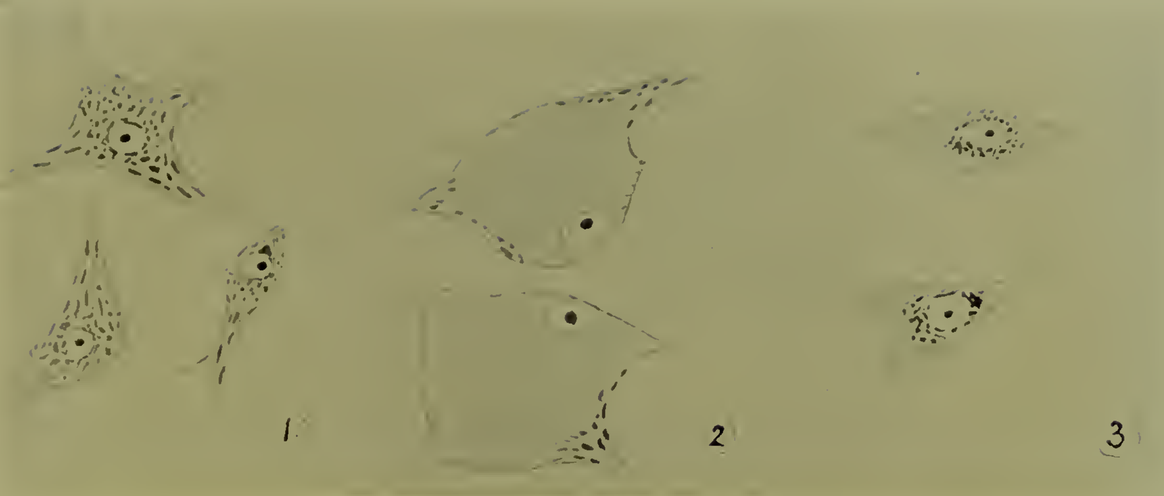
V. *Marchi's stain.* Degenerated fibres in anterior pyramidal tract of spinal cord, stained deep black, to the left side of the figure. In the middle of the figure is the anterior median fissure. To the right is the normal anterior pyramidal tract, in which no degenerated fibres can be seen.

VI. Nitrate of silver and formol stain (*Bielschowsky's method*). Transverse section of spinal white matter; axis cylinders black.

VII. *Bielschowsky's method.* Anterior grey matter. Nerve cells and fine fibres black.

VIII. *Bielschowsky's method.* Longitudinal section of spinal white matter; axis cylinders black.

I.



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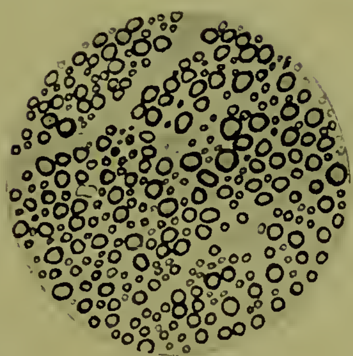

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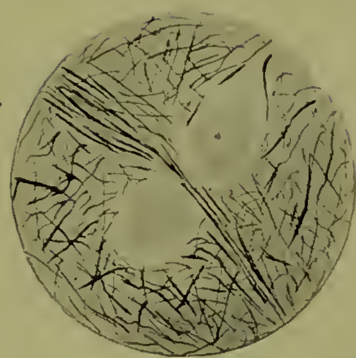

IV.

V.

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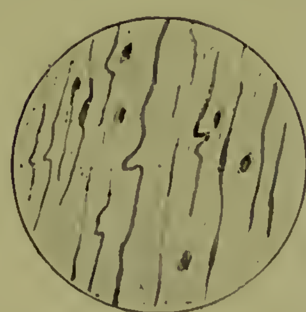

PLATE I.

in most instances it appears like a straight piece of black thread on a white or yellowish ground (in section stained according to Golgi's method). As regards the axis-cylinder processes, there are two forms of nerve cells :—

1. Cells with a very long axis-cylinder process, which becomes ultimately the axis-cylinder of a central or peripheral nerve fibre. In these cells the axis-cylinder process has numerous offshoots or collaterals.

2. Cells with a short axis-cylinder process which divides and subdivides into numerous small branches. Such processes do not become the axis-cylinders of nerve fibres.

The collaterals of the axons of the first variety of cells form most of the fine nerve fibres, seen in the grey matter of the cord, in section stained according to Weigert's method.

The *internal structure* of nerve cells can be well studied by the staining of sections according to Nissl's method, or with toluidin-blue. When stained according to these methods the nuclei and especially the nucleoli, and, in many cells, granules in the protoplasm are stained deep blue. According to their reaction to this stain nerve cells may be divided into two forms :—(1) Cells in which both the nucleus and granules in the protoplasm are stained — *Somatochrome* cells. The large ganglion cells of the anterior horns of grey matter of the spinal cord are of this variety. (2) Cells in which the nucleus only is stained by Nissl's method, or its modifications—*Caryochrome* cells. The protoplasm of these cells is unstained. This form of cell is met with in the posterior horns of the cord and in the gelatinous substance of Rolando.

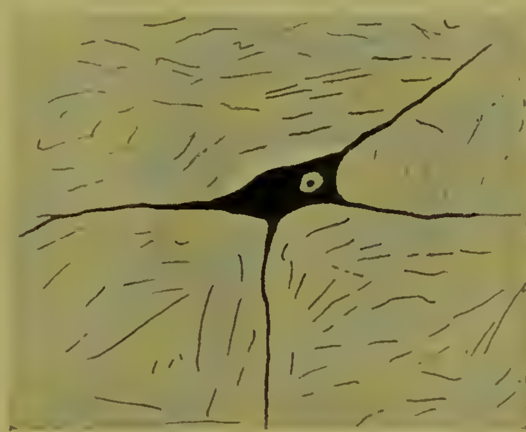

FIG. 5.—Nerve Cell of Anterior Horn of Spinal Cord. Formol and silver nitrate method (writer's modification of method).

In the somatochrome cells (such as the nerve cells of the anterior horns of grey matter of the spinal cord) when stained according to Nissl's method or its modifications, the protoplasm is seen to consist of two parts : one portion which is deeply stained, another which is unstained. To the stained portions of the cell protoplasm are given the names of *chromophile* or *tigroid* substance, or Nissl's granules. This stained part of the cell consists of granules, irregularly shaped blocks, short rods, spindle-formed and triangular masses. The stained or tigroid substance is found both in the protoplasm of the cells and also in the cell processes or dendrites ; but it is not found in the axis-cylinder process. In many nerve cells, and especially in those of the cerebro-spinal ganglia, the stained or chromophile substance is absent in the part of the cell from which the axis-cylinder arises. There is, at this region, a clear triangular area free from stained granules—the cone of origin of the axis-

cylinder. The chromophile substance is also absent in the fine *branches* of the protoplasmic cell processes. In some cells, especially in the large cells of the spinal ganglia of the posterior nerve roots, there is a narrow zone at the periphery of the cell, and also one just around the nucleus, in which the chromophile substance is absent. As regards the different forms of stained granules, the rods and spindles are found at the periphery of the cells and in their large protoplasmic processes. Irregular blocks of chromophile substance have sometimes a concentric arrangement. In spindle-shaped nerve cells, at each pole of the cell, there is often a large triangular block of stained substance, with one side hollowed out, which sits like a cap on the nucleus. At the point of division of the protoplasmic processes there are often triangular masses of chromophile substance. In some cells the stained granules are irregular in shape and without any definite arrangement (*see* Fig. 4, cell to the right, and Plate I).

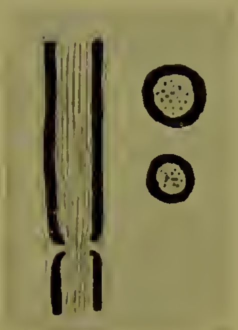

FIG. 6.—Fibrillæ in Axis-cylinder of Nerve Fibres. Longitudinal and transverse sections (diagrammatic).

The unstained part of the cell protoplasm —chromophobe part—consists (according to many observers) of a fine network, immersed in a homogeneous substance. Various points of this network are incrusted with the stained granules and blocks already described. Bethe, Apathy and others have described fibrillæ in the cell protoplasm, which are quite independent of each other, and can be followed for a long distance. These fibrillæ are seen in the axis-cylinder process, and also in the dendrites ; when they reach the cell protoplasm they diverge and run through the body of the cell to other processes or to the axis-cylinder. The fibrillæ in the axis-cylinder and nerve cells are seen beautifully in sections stained according to the recent methods of Bielchowsky and Ramon y Cajal[1] (*see* Fig. 4, central cell, and Fig. 6).

In many nerve cells yellowish pigment granules are seen. These increase with the age of the individual. The granules stain black both with osmic acid and in Marchi's method of staining. They are therefore probably fatty in nature, but they do not dissolve in ether or alcohol.

The *nucleus* of a nerve cell has a globular appearance ; it has a distinct membrane separating it from the cell protoplasm. In the centre of the nucleus is a large nucleolus, which stains deeply. Usually it is single, and from it irregular fine trabeculae pass to the membrane of the nucleus.

The nerve cells are surrounded by a peri-cellular space, the walls of which sometimes possess endothelial cells. The cells of the spinal ganglia are surrounded by a connective tissue capsule which is lined by endothelial cells on its inner side.

Neuron Theory.—On the observations of Golgi, Forel, Ramon y Cajal,

[1] *See* Donaggio's drawings, *Review of Neurology and Psychiatry*, No. 2, 1905.

His and Waldeyer is based the neuron theory. Though there are now certain objections to this theory, it is still useful in enabling a right conception to be formed of the structure of the nervous system. According to the neuron theory, the processes of the nerve cells, both axons and dendrites end in a network of fibres, but the endings of all are free ; they may cross and interlace, but are not directly united with the processes of other cells or with other nerve fibres. To a ganglion cell with all its processes the term neuron is applied.

The whole nervous system is built up of neurons (cells and their processes) held together by a supporting substance, and according to the neuron theory one neuron comes into relation with one or more adjacent neurons by contact only. Recent observations, however, have shown that in the axons are fine fibres—fibrillæ—which pass into the ganglion cells, run through its protoplasm and pass into the dendrites. Some observers believe that by these fibrillæ the continuity of the whole nervous system is established.

Tracts of Fibres in the White Substance of the Spinal Cord.

The white substance of each half of the spinal cord is divided into two parts : (1) the posterior columns—between the posterior horn of grey matter and the posterior median septum ; and (2) the antero-lateral region, occupying the lateral part of the cord (on the outer side of the posterior and median grey matter), surrounding the anterior horn, and extending to the anterior median fissure.

These areas of white matter have been subdivided into a number of tracts, which have been mapped out, (1) by the study of the embryology of the spinal cord, and (2) by the study of the degeneration of spinal nerve fibres after pathological lesions in man or experimental lesions in animals.

The study of the development of the spinal cord of the foetus has shown, that various bundles of fibres acquire their myelin sheaths at different periods. Flechsig and others have differentiated numerous tracts of fibres by this method. Thus, at birth, the fibres of a tract in the lateral white matter, and of another in the anterior white matter close to the anterior median fissure, do not possess a medullary sheath ; whilst the surrounding fibres of the white matter possess a normal medullary sheath. Hence, in sections stained according to Weigert's method, the tracts just mentioned appear paler than the rest of the white matter. These tracts are the crossed and direct pyramidal tracts.

The second method of differentiating the tracts of nerve fibres—by the study of the degeneration following various pathological lesions—is based on the fact that spinal nerve fibres degenerate when separated from their cells of origin, just as peripheral nerve fibres degenerate on the peripheral side of the lesion, when they are separated from their cells of origin in the anterior horn of the spinal cord. This method of investigation

was studied by Türck long ago (*see* Fig. 33). In the first method of study (developmental) Weigert's stain is of great service, in the second method Marchi's stain is particularly useful.

The studies of Türck, Flechsig, Gowers and numerous other observers

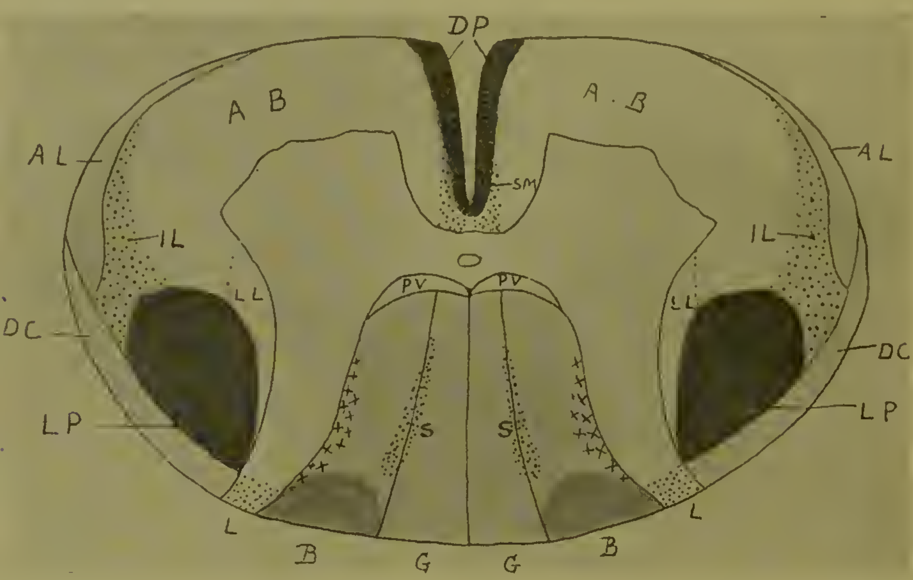

Fig. 7.—Tracts of Fibres in the White Matter of the Cord (diagrammatic).

Posterior Columns.

G = Posterior median—Goll's column.
B = Posterior external—Burdach's column.
S = Comma-shaped tract of Schultze (dotted) (Fig. 7), also Fig. 8 (1).
Hoche's bundle (mid. and lower dorsal region), Fig. 8 (2), on posterior surface of cord.
Oval field of Flechsig (lumbar region) Fig. 8 (3).
Triangle of Gombault-Philippe (sacral region), Fig. 8 (4).
Posterior root zone marked with x x (Fig. 7).
P V = Posterior ventral field.

Antero-lateral Columns.

L P = Crossed or lateral pyramidal tracts.
D C = Direct cerebellar tract.
A L = Ascending antero-lateral tract of Gowers.
D P = Direct pyramidal tract.
S M = Ascending sulco-marginal tract of Löwenthal and Marie (dotted).
Descending sulco-marginal fibres in same region.
I L = Intermedio-lateral tract of Löwenthal (Rubro-spinal tract of Monakow in same region).
L L = Lateral limiting layer.
L = Lissauer's zone (dotted).
H = Helweg's bundle (Fig. 9, cervical region).
A B = Anterior ground bundle: region of endogenous fibres; of descending antero-lateral tract (*see* Fig. 10); and of fibres from cerebellum, Deiter's nucleus, the corpora quadrigemina, pons, and sensory fibres (spino-thalamic, passing upwards to the brain).

have enabled us to differentiate many tracts in the white matter of the cord. The following are the more important.

The *posterior columns* consist of (1) the posterior median columns or columns of Goll—close to the posterior median septum, and (2) the posterior external columns of Burdach on the outer side of the latter. The two

are separated, at least in the cervical region, by the posterior intermediate septum. In both of these tracts most of the fibres have an ascending course. In addition, in the posterior columns there is a comma-shaped tract in the upper part of the cord between the columns of Goll and Burdach—a tract of descending fibres, the comma-shaped tract of Schultze. Of less importance are the following tracts of descending fibres in the posterior columns. On the posterior surface of the cord near the posterior median septum, in the middle and lower dorsal region, is a small bundle of fibres, the position of which is indicated in Fig. 8 (2), Hoche's bundle. In the lumbar region there is an oval-shaped bundle close to the median septum, shown in Fig. 8 (3), the oval field of Flechsig. In the sacral region there is a triangular bundle, near the median septum, having the position shown in Fig. 8 (4), the triangle of Gombault and Philippe.

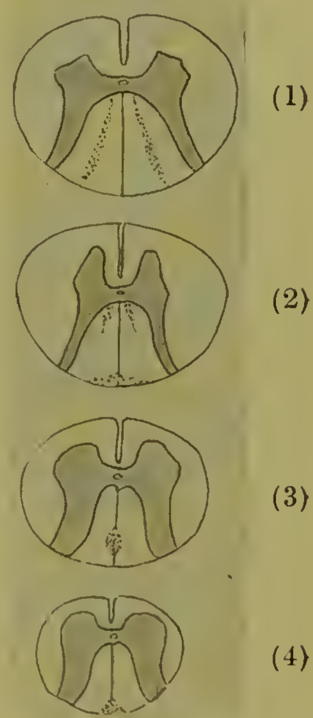

Fig. 8. — Showing Descending Fibres in Posterior Columns. (1) Comma - shaped tract of Schultze. (2) Hoche's bundle on posterior surface of cord. (3) Flechsig's oval field. (4) Triangle of Gombault-Philippe.

To the descending fibres in the lumbo-sacral region of the cord, Bruce and Muir have given the name of descending septo-marginal tract. Obersteiner has named the descending fibres the dorso-medial bundle.

Near the posterior commissure is a strip of fibres in the position marked P V in Fig. 7—the ventral posterior field. Close to the posterior horn (on its median side) is the region at which numerous posterior root fibres pass into the grey matter—posterior root (entrance) zone. In the cervical region the dorsal part of Burdach's column has been described as the posterior external field.

At the tip of the posterior horn are the fine fibres forming Lissauer's bundle.

In the *antero-lateral columns* are the following tracts : (1) The crossed or lateral pyramidal tracts (L P). In the lower cervical and dorsal regions this tract does not extend to the surface of the cord, but in the lumbar region it reaches the surface. (2) On the surface of the cord is the direct or dorsal cerebellar tract in the cervical and dorsal regions. (3) In front of this latter tract, and also on the surface of the cord, is the ascending antero-lateral tract of Gowers, or ventral cerebellar tract. (4) In the anterior columns close to the anterior median fissure is the direct pyramidal tract of Türck. (5) Between the crossed or lateral pyramidal tract and the outer side of the grey matter is the lateral limiting layer.

Other tracts of less importance have also been differentiated. On each side of the anterior fissure are fibres forming the ascending sulco-marginal tract of Löwenthal and Marie, and in the same part are the

fibres of the descending sulco-marginal tract ; also mixed with the fibres of the direct pyramidal tract are fibres from the cerebellum.

In front of the crossed pyramidal tract is the intermedio-lateral tract of Löwenthal (I L). Mixed with the fibres of this tract are long fibres which come from the red nucleus behind the optic thalamus : to these fibres the name of Monakow's tract is given.

On the surface of the cord, in the upper cervical region (indicated in

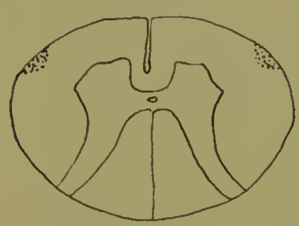

Fig. 9.—Helweg's Bundle : Cervical Region.

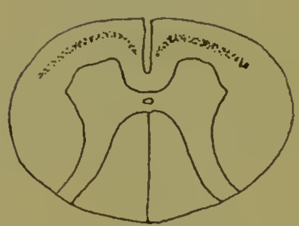

Fig. 10. — Descending Antero-lateral Tract.

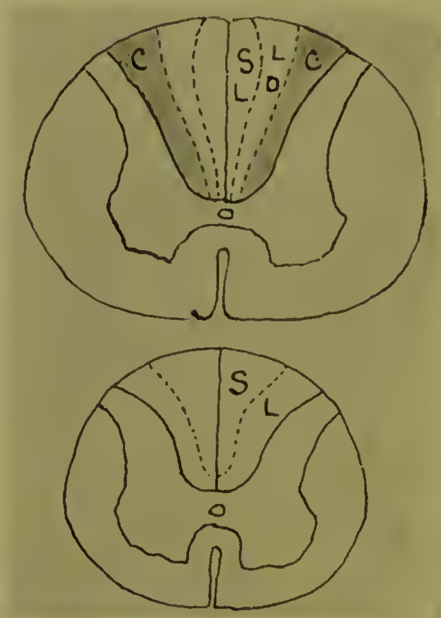

Diagram showing Position of Fibres of Posterior Columns.
Fig. 11 (lower figure.) = Lumbar region.
L = Lumbar root fibres.
S = Sacral posterior root fibres.
Fig. 12 (upper figure). = Cervical region.
C = Fibres from arm and cervical region.
S L = Fibres of posterior nerve roots of sacro-lumbar region.
L D = Fibres from upper lumbar and dorsal nerve roots.

Fig. 9) is a small triangular tract of fibres, Helweg's bundle. These fibres have been traced upwards to the olivary body (Edinger) and downwards to the third cervical segment.

To the white fibres around the anterior horn, not included in the tracts named, the term anterior ground bundle has been applied.

In the antero-lateral region near the surface of the cord, a tract the position of which is indicated in Fig.10, has been described by many observers as the antero-lateral descending tract. It contains fibres from Deiter's nucleus, from the dentate nucleus of the cerebellum, and from the corpora quadrigemina.

The region around the anterior horn, marked A B in Fig. 7 contains the fibres of many tracts mixed together—(1) the antero-lateral descending tract, descending fibres from the cerebellum, Deiter's nucleus,

corpora quadrigemina and pons; (2) ascending fibres, spino-thalamic, which arise in the grey matter of the opposite side of the cord, and run upwards to the brain, conveying, in all probability, sensory impulses; (3) association fibres (endogenous) which link together, or connect, various segments of the spinal cord.

Figs. 11 and 12 show how the fibres from the posterior roots, which enter the posterior columns at the lower part of the cord, gradually pass toward the median septum. In the lumbar region the fibres from the sacral nerve root are situated close to the median septum: those from the lumbar roots are near the posterior horns. In the cervical region the sacral and lumbar fibres are adjacent to the posterior septum: the fibres from the cervical root are near the posterior horns: the dorsal fibres occupy an intermediate position (Fig 12).

* * * * *

After this description of the tracts of fibres seen in transverse sections of the spinal cord we may consider the tracts with reference to their origin.

It is important to remember that the fibres entering and leaving the cord in the spinal nerve roots are much more numerous than those passing to and from the brain at the upper cervical region.

The fibres in the white matter of the cord may be divided into three groups: (1) Fibres which pass from one level of grey matter to another level. These fibres link together the grey matter of adjacent or more distant spinal segments. They are the endogenous, intrinsic, or association fibres. (2) Fibres which pass from the brain to the cord,—fibres arising from nerve cells in the brain. (3) Fibres which pass from the cord to the brain,—fibres arising from nerve cells in the cord or posterior ganglia. Groups 2 and 3 form the exogenous or extrinsic fibres.

As regards the *endogenous* or association fibres which pass from one segment of the cord to another, the shorter of these connect adjacent segments and lie close to the grey matter, the longer connecting together more distant segments are situated farther away from the grey substance. These endogenous fibres are situated chiefly in the posterior ventral field, the lateral limiting layer, sulco-marginal tract, and scattered throughout the antero-lateral region (anterior ground bundle).

As regards the *exogenous* fibres, the following are the most important of the tracts passing from the brain to the spinal cord:—

1. The cortico-spinal or pyramidal tracts, crossed and direct, from the motor region of the brain cortex. They convey motor impulses from the brain, and finally terminate in the neighbourhood of the cells of origin of the anterior nerve roots. (According to some histologists they are connected with the cells of the anterior horns by a second neuron.)

2. The rubro-spinal tract, which has been studied by Monakow, Collier, F. Buzzard and many others. It is situated just in front, and to the outer side, of the crossed pyramidal tract. Fibres pass downwards in this tract from the red nucleus (just behind the optic thalamus). The fibres finally pass to the anterior horns of spinal grey matter.

3. Near the surface of the cord, in the antero-lateral region, a tract of fibres, the antero-lateral descending tract, has been described by many observers, in which fibres pass down from various parts of the brain—from Deiter's nucleus, the dentate nucleus of the cerebellum, and the corpora quadrigemina.

Many tracts of minor importance have been described in this antero-lateral region. (*See* writings of Edinger, Collier, F. Buzzard, and others.)

Descending fibres in the posterior columns pass downwards in the comma-shaped tract of Schultze, the oval area of Flechsig, the septo-marginal tract (of Bruce, Muir, and Hoche), and the sacral triangle of Gombault and Philippe. The origin of these fibres has not been definitely decided.

Fig. 13.—Tracts conveying Impulses *from* the Brain. (1) Crossed (lateral) and direct pyramidal tracts. (2) Rubro-spinal tract (intermedio-lateral tract in same region). (3) Antero-lateral descending.

Fig. 14a.—Tracts conveying Impulses *to* the Brain. (1) Direct cerebellar. (2) Antero-lateral ascending tract of Gowers. (3) Spino-thalamic in lateral and posterior columns.

The following are the chief tracts conveying impulses to the brain :—

1. The lateral or direct cerebellar tracts, conveying fibres from the cells of Clarke's column to the worm of the cerebellum through its lower peduncles (dorsal spino-cerebellar tract).

2. The tract of Gowers (antero-lateral ascending or ventral spino-cerebellar) conveying fibres to the worm of the cerebellum through its upper peduncles.

3. The spino-thalamic tracts which convey impulses from the posterior nerve roots to the optic thalamus. These fibres run upwards in the posterior columns and also in the lateral columns (*see* Fig. 14a).

A large portion of the posterior root fibres enter the grey matter of the posterior horns soon after passing into the cord. They divide into fine branches around the cells of the posterior horns ; from these cells fibres arise which form a secondary sensory neuron or tract. These fibres pass to the anterior commissure, then cross to the anterior and lateral columns of the opposite side of the cord and pass up to the optic thalamus. Other fibres of the posterior root pass upwards in the posterior columns, without crossing, to the nuclei in the medulla. Fibres from the cells of these nuclei (to which the sensory impulses are

conducted) cross later and probably pass to the optic thalamus. Thus the whole of the sensory tracts convey impulses to the opposite side of the brain (*see* Fig. 14B).

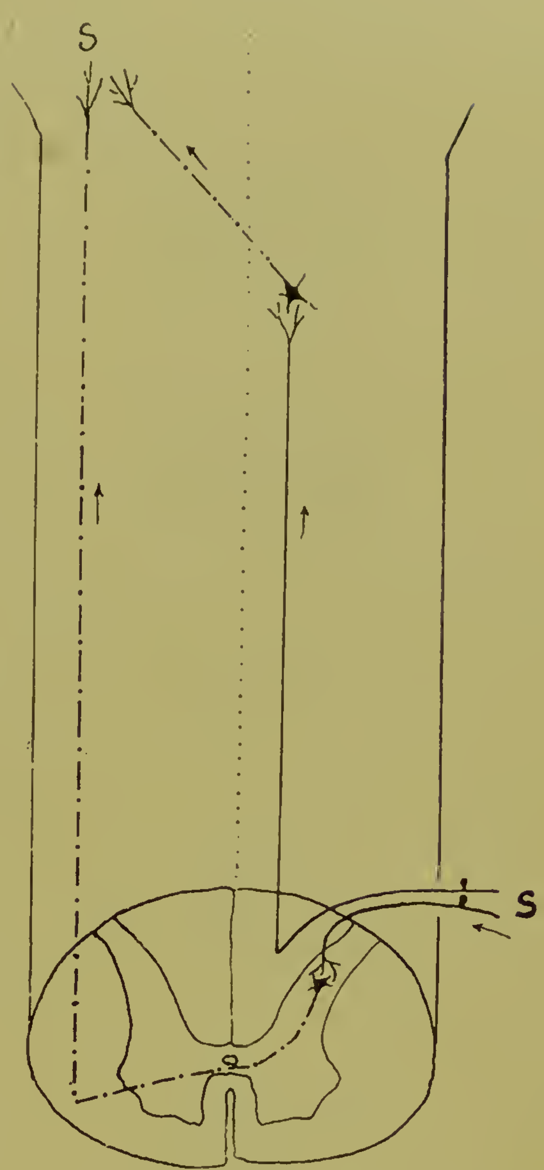

FIG. 14B.—Showing Course of Fibres of the Posterior Nerve Root. Paths for impulses which ascend without crossing in the Spinal Cord, and paths for impulses which cross in the Spinal Cord.

THE COURSE AND ANATOMICAL RELATIONS OF THE CHIEF TRACTS OF THE CORD.

The crossed or lateral pyramidal tract, which contains fibres coming from the motor area of the cerebral cortex, extends down to the lowest part of the cord (*see* Fig. 15). At its anterior part it is separated from the grey matter by a zone of fibres, the lateral limiting layer, which is probably composed of commissural fibres connecting together different levels of the grey matter. The crossed or lateral pyramidal tract is roughly triangular in shape. It is separated from the external surface of the cord in the cervical and dorsal regions by the direct cerebellar tract. In the lumbar region the crossed pyramidal tract is situated just under the pia mater, on the surface of the cord. Also in the upper cervical region (third cervical segment) the tract comes to the surface of the cord (*see* Fig. 15).

The direct pyramidal tract of Türck is usually stated to end at the mid-dorsal region; but it may extend as far as the lumbar region or to the lower end of the cord (Russell). Most of its fibres pass ultimately through the anterior commissure to the grey matter of the opposite side.

Most of the fibres of the crossed pyramidal tract come from the nerve cells of the motor cortex of the opposite side of the brain, but a few fibres come from the brain cortex of the same side. The fibres of the direct pyramidal tract come from the nerve cells of the motor cortex of the brain on the same side. Thus most of the fibres from the nerve cells of the motor cortex decussate in the medulla and pass into the lateral pyramidal tract of the opposite side of the spinal cord, other fibres do not decussate, but pass into the direct pyramidal tract, and a few into the

lateral pyramidal tract, on the same side as the brain cortex from which they arise. The fibres of both tracts ultimately pass to the grey matter of the anterior horn. Here they divide and their processes come into relation with the processes of nerve cells of the anterior horn (*see* note on page 18). One process of a nerve cell of the anterior horn becomes the axis-cylinder of a peripheral nerve.

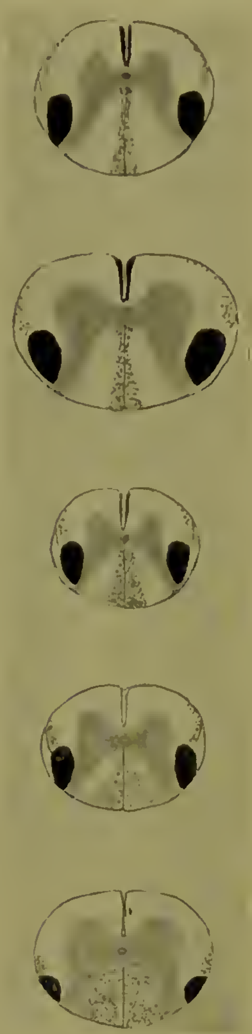

FIG. 15.—Sections showing relative positions of Tracts in the Cord. Crossed and direct Pyramidal Tracts = deep black. The direct cerebellar tracts on the lateral surface of the Cord = clear. In front of this tract is the antero-lateral ascending tract of Gowers = dotted. Fibres from the posterior roots in the lumbar region (lowest figure) = dotted area. These fibres gradually pass into the posterior median column of the cervical region.

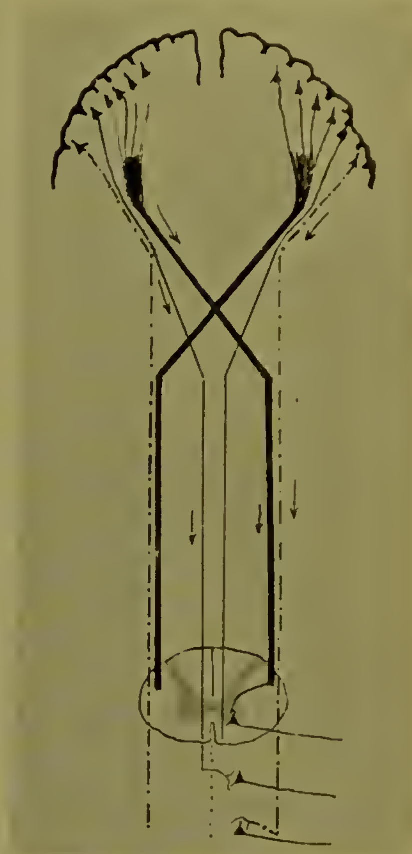

FIG. 16.—Motor Fibres from the Brain Cortex. Fibres of lateral or crossed pyramidal tracts = deep black line. Fibres of direct pyramidal tracts = fine line. Fibres of lateral pyramidal tracts which have not decussated = interrupted line.

Thus we have the upper and lower motor neurons—the motor cell of the brain cortex and its long process being the upper neuron, the cell in the anterior horn and its process in a peripheral nerve the lower neuron (*see* Figs. 16 and 17). The function of the pyramidal tracts, crossed and direct, is to convey motor impulses from the brain cortex.

The intermedio-lateral bundle degenerates below a transverse cord lesion, and the fibres of this tract are said to descend from the red nucleus

behind the optic thalamus. Many of the fibres of the anterior ground bundle and of the lateral limiting layer are probably commissural fibres and connect different levels of the grey matter of the cord (*see* p. 21).

The fibres of the posterior median column of Goll pass upwards to the medulla and end in the nucleus gracilis. Many fibres of the posterior external column of Burdach pass into the posterior median column, but some run up to the cuneate nucleus of the medulla. At the upper part of the cord, the fibres of the posterior median column are situated more posteriorly the lower their origin, as already described (*see* p. 13 and Figs. 11 and 12).

The fibres of the two posterior columns are afferent, and come from the posterior nerve roots. The fibres of the posterior roots that enter the posterior median columns do not decussate.

The direct cerebellar tract ends at the first lumbar nerve root. In the cervical and dorsal regions it separates the crossed pyramidal tracts from the surface of the cord. At the highest cervical region (third segment) the tract occupies a position a little further forward, and the posterior part of the crossed pyramidal tract comes to the surface of the cord.

The direct cerebellar tract receives its nerve fibres from the cells of the column of Clarke and the tract only degenerates when the lesion is at or above its lower origin, i.e. junction of the dorsal and lumbar regions of the cord. There is no degeneration of the tract in a lesion below this level.

The antero-lateral ascending tract of Gowers commences at the lower end of the cord. Hence a lesion of the middle lumbar region of the cord causes degeneration of the antero-lateral ascending tract of Gowers, but no degeneration of the direct cerebellar tract. The direct cerebellar tract and the ascending antero-lateral tract of Gowers both end in the cerebellum.

The tract of Gowers probably conveys impulses upwards from the opposite posterior nerve roots.

Lissauer's tract contains fibres from the posterior roots which course upwards for a short distance and then enter the posterior horn.

Spinal Neurons.

As already mentioned the fibres of the spinal cord are of two kinds: (1) *endogenous*, or those which arise from nerve cells within the cord and pass from one level of the cord to another, (2) *exogenous*, or those which arise from cells outside the cord.

Of the *exogenous* motor fibres those of the pyramidal tracts are the most important. They arise from the motor ganglion cells of the cortex of the brain, pass down through the internal capsule, crus, pons and medulla, and then enter the crossed and the direct pyramidal tracts. The cells and fibres just described form the upper motor neurons.

The fibres of the lateral or crossed pyramidal tracts, as well as fine side branches—collaterals—which arise from them, pass finally into the

anterior horn of grey matter of the same side and divide into a fine network. The fibres of this network come in contact with the processes of the nerve cells of the anterior horn of grey matter. (According to some histologists fibres from the crossed pyramidal tract pass into the posterior grey matter and are then connected by a secondary neuron in the grey matter with the motor cells in the anterior horn (*see* Figs. 17 and 18).

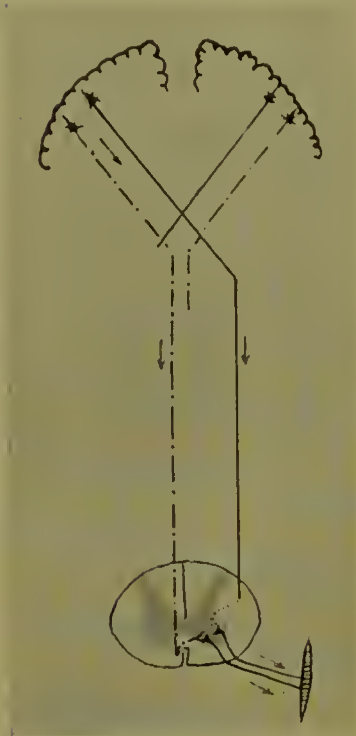

Fig. 17.—Upper and Lower Motor Neurons. Fibres in the lateral pyramidal tract = unbroken line. Fibres in the direct pyramidal tract = broken line.

The nerve cells of the anterior horns together with the nerve fibres of the anterior nerve roots form the lower motor neurons.

The posterior columns of the spinal cord are composed of (1) exogenous fibres, entering the cord from the posterior nerve roots, and (2) of endogenous fibres, which arise in the cord itself and pass from one level to another.

The exogenous fibres coming from posterior nerve roots have their cells of origin in the ganglia of this root. (Possibly the cells of origin of a few posterior root fibres are in connexion with the peripheral sensory nerves.) The cells of the posterior root ganglia appear unipolar in the adult, because the two processes lie close together for a short distance (*see* Fig. 19). One process passes to a peripheral nerve, the other enters the cord in the posterior nerve root.

The posterior root fibres consist of a lateral and a median bundle. The lateral fibres pass into Lissauer's zone, and finally end around cells in the posterior horn. The fibres of the median bundle pass into Burdach's column.

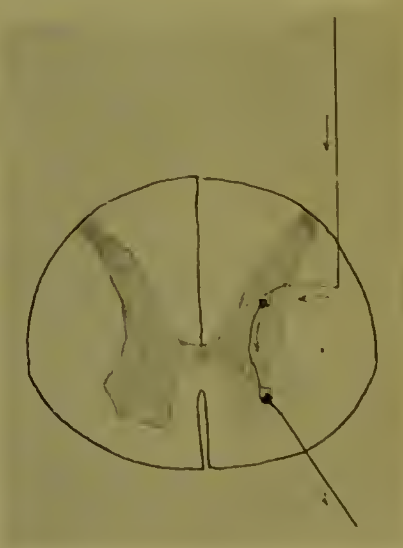

Fig. 18. — Upper and Lower Motor Neurons: secondary neuron in grey matter.

The posterior root fibres divide, in a Y-shaped manner, into two branches, directly after entering the cord. One branch is ascending, the other is descending. The latter soon enters the grey matter and divides into numerous branches. Descending fibres pass into the comma-shaped tract of Schultze.

Some of the fibres of the posterior root enter the grey matter of the posterior horn directly, others enter after running upwards for a distance, other fibres run up to medulla.

The descending fibres are mostly short. The ascending fibres consist of both short and long fibres : the short ascend for a little distance then bend at a right angle and pass into the grey substance. The long fibres run up to the medulla, in the posterior columns, and end in the grey substance of the medulla—nucleus gracilis and nucleus cuneatus. All fibres of the posterior roots which do not pass up to the brain in the posterior columns soon end in the grey matter. All the fibres in the posterior columns,

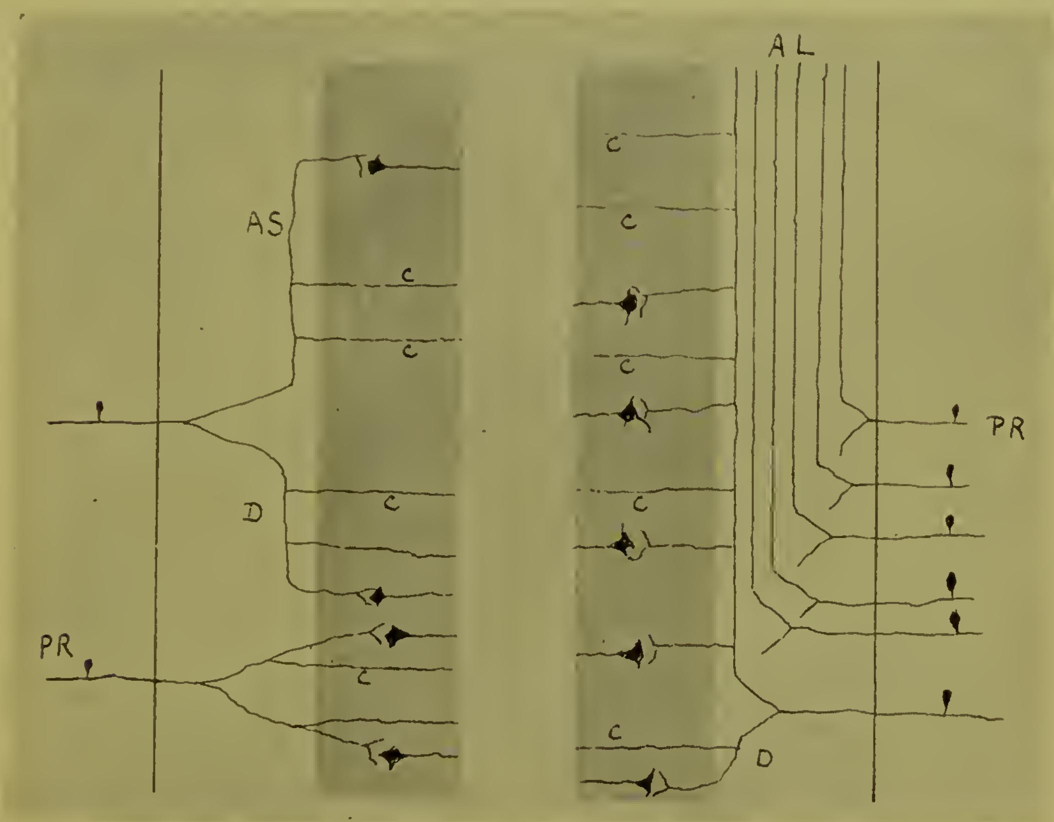

FIG. 19.—Course of the Fibres of the Posterior Nerve Roots in a Longitudinal Section of the Cord (diagrammatic).

PR = Posterior root fibres which divide in a **Y**-shaped manner on entering the Cord.
AL = Long ascending fibres.
D = Descending fibres.
AS = Short ascending fibres.
C = Collaterals.
Deeply shaded part = grey matter.

both ascending and descending, give off collaterals which pass into the grey matter, especially at the middle third of Burdach's column. Hence all the descending fibres, all short ascending fibres, and all collaterals end in the grey matter of the spinal cord.

The fibres entering the grey matter end in fine branches around the cells of the grey matter. (i) They end mostly around the nerve cells of the posterior horn and around the middle zone cells on the same side (the former only are indicated in diagram). (ii) Some fibres end around the motor cells of the anterior horn of the same side. (These fibres are

collaterals which probably convey reflex impulses—reflex collaterals.) (iii) Other fibres end around the cells of the column of Clark; (iv) A small number of collaterals cross in the posterior commissure to the opposite side of the cord and pass into the posterior horn.

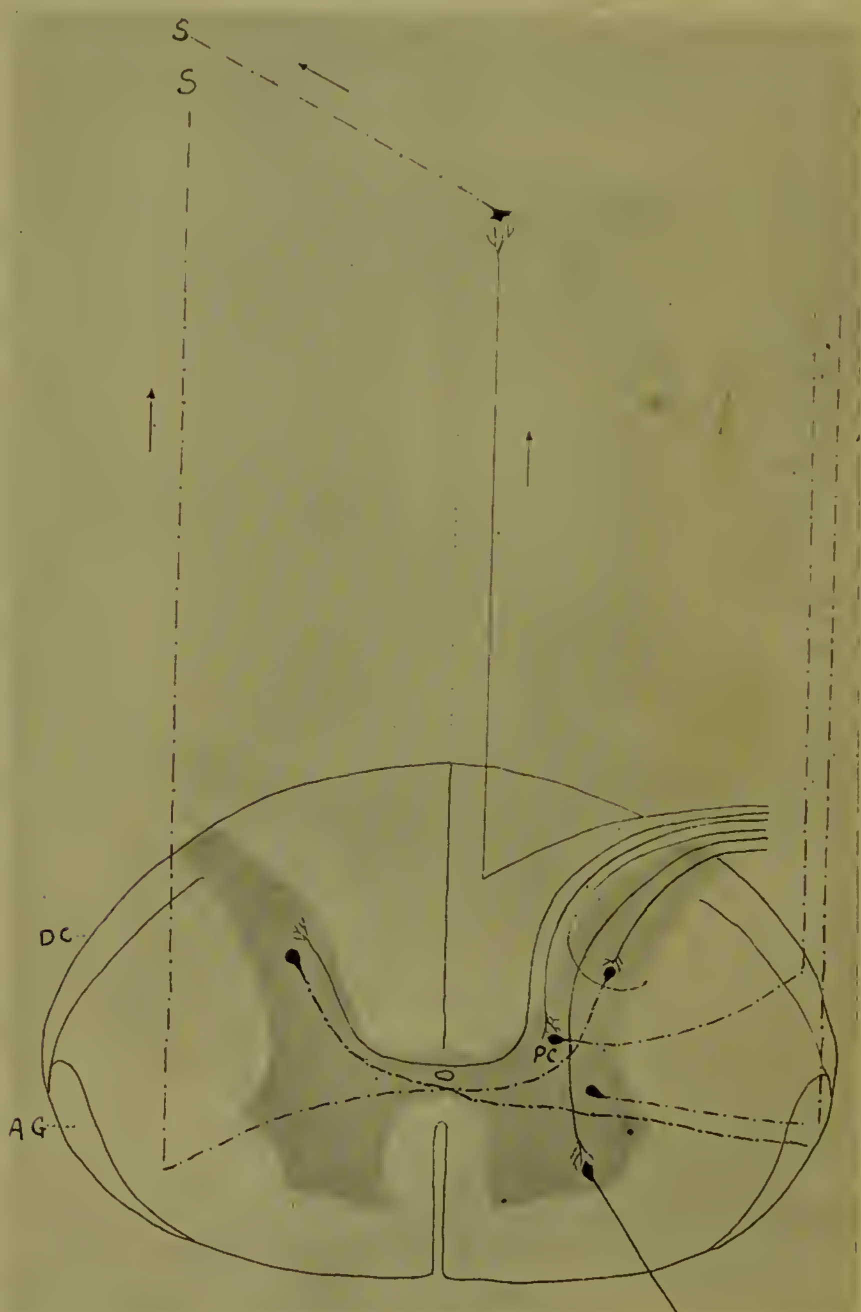

Fig. 20.—Sensory Neurons. Course of fibres from posterior roots in transverse section of Cord. DC = Direct cerebellar tract. AG = Ascending antero-lateral tract of Gowers. PC = Column of Clarke. Interrupted lines = fibres from cells in the grey matter (secondary sensory neurons). At the top of the figure the oblique line S indicates a neuron in the medulla conveying impulses, from the uncrossed fibres of the posterior column, to the opposite side of the brain.

The nerve cells of the grey substance of the posterior horns and median grey substance and their processes form secondary neurons. The axis-

cylinder processes of these cells probably pass to the anterior commissure, and cross to the opposite antero-lateral column and ascend to the optic thalamus.

Some of the fibres of the posterior root pass into the lateral limiting layer: their further course is not known.

The posterior column consists chiefly of exogenous fibres; only a small portion are endogenous. The long ascending fibres of the posterior roots, in their upward course, pass from the lateral to the median part of the posterior columns. Fibres which enter the lowest part of the cord have the longest course and are situated, at the upper cervical region, at the posterior and median region of Goll's columns.

From the cells of Clarke's column fibres pass into the lateral cerebellar

Fig. 21.—Endogenous Association Neurons, 1, 2, 3, 4. A = Motor neuron of anterior horn and nerve root. Interrupted lines = other endogenous neurons (modified after Dejerine). The region occupied by commissural and association fibres in the white matter is shaded.

tract and run up to the cerebellum ending in the superior vermiform process.

The fibres of the ascending antero-lateral tract of Gowers probably come from the nerve cells of the intermediate grey matter (or posterior horn) of the same or opposite side.

Fibres from the nerve cells of the anterior horn of grey matter pass into the anterior nerve root.

In addition to the cells already mentioned, the grey matter contains numerous cells which have a different function (association, commissural or endogenous fibres). Fibres from some of the cells in the median region of the anterior horn fibres pass into the anterior commissure, then cross to the other side of the cord, enter the anterior column and run in a longitudinal direction giving off processes to the grey substance. Fibres

from other scattered cells of the grey matter pass into the lateral and anterior columns, divide into ascending and descending branches, which give off side branches—collaterals—to the grey matter. These are commissural fibres which connect together different levels of grey matter. The posterior columns also contain commissural fibres in the posterior ventral fields. The endogenous and commissural fibres of the cord are situated close to the grey matter, both anteriorly and posteriorly.

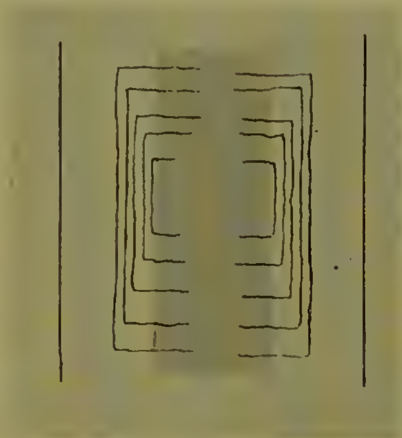

FIG. 22. — Diagram of Longitudinal Section of Cord. Grey matter deeply shaded. Commissural or association fibres linking together different levels of grey matter.

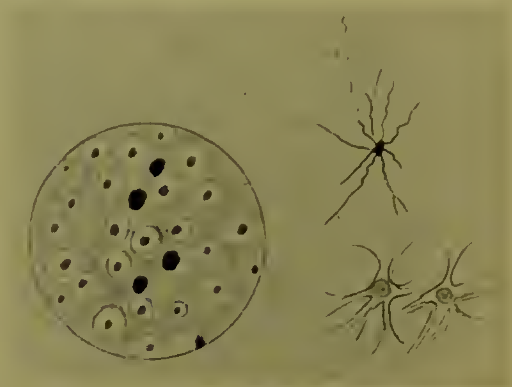

FIG. 23.—Neuroglia Cells. The section within the circle shows neuroglia cells in transverse section of white matter (logwood and eosin). The nuclei of the cells are deep black (four in section). To the right of the figure at the upper part is a neuroglia cell stained by Golgi's method. In the lower part are two cells stained by Weigert's neuroglia method.

Fig. 21 represents some of the more important sources of origin of these commissural fibres.

The comma-shaped tracts of the posterior columns are said to contain descending fibres from the posterior roots.

THE NEUROGLIA.

Between the white fibres, as already mentioned, is the connective tissue substance—the neuroglia. This tissue contains fine fibres and small cells with a round nucleus ; but the cell protoplasm is so small that only the nucleus is seen by a low power of the microscope.

The finer structure of the neuroglia has been demonstrated by two methods—Golgi's stain and Weigert's neuroglia stain. In the sections stained according to Golgi's method the neuroglia is seen to contain cells with numerous branching processes. In sections stained according to Weigert's neuroglia method, the neuroglia fibres are smooth and show no branches or processes, they run by or through the body of the neuroglia cell, or only lie in contact with it. The neuroglia fibres, by this stain, appear therefore as true intercellular substance.

According to Lugaro the results obtained by these two stains are not

really contradictory. In the embryo the neuroglia is composed of cells with protoplasmic processes, but no intercellular substance, i.e. no neuroglia fibres. When these cells are fully developed some of them send out or excrete neuroglia fibres. Then the cell undergoes a kind of regressive change. The nucleus becomes smaller and darker, the protoplasmic processes and the cell body atrophy.

Both in Golgi's and Weigert's preparations, neuroglia fibres can often be seen which are independent of the cell body.

In the adult different forms of neuroglia cells are seen: cells with

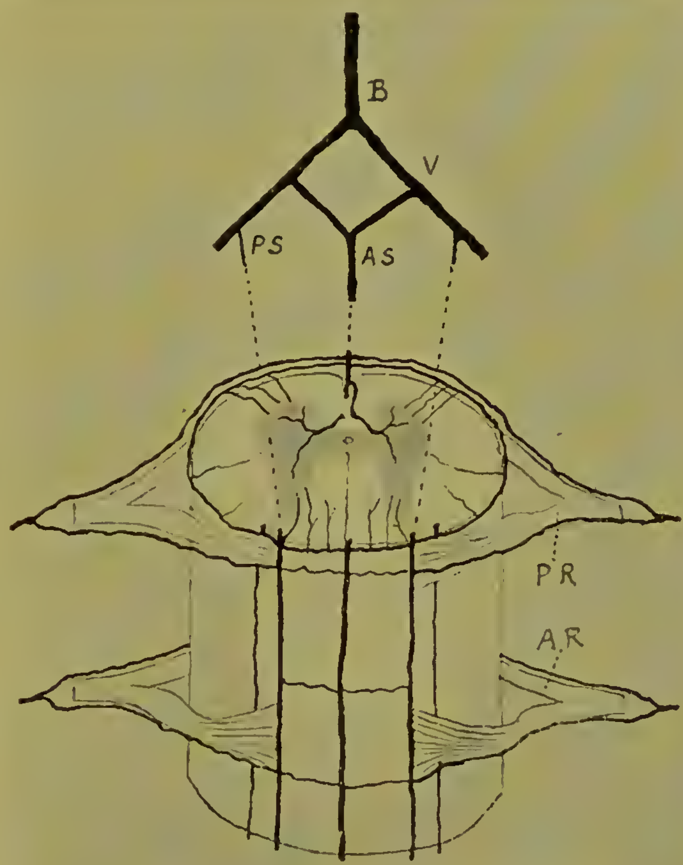

FIG. 24.—Arteries of the Spinal Cord (modified after van Gehuchten and others).

AS = Anterior spinal artery arising from the vertebrals at the medulla.
PS = Posterior spinal.
V = Vertebral.
B = Basilar.

Note.—Circle of arteries and vascular anastomosis in the pia mater; also the small arteries running along the anterior and posterior nerve roots AR and PR.

smooth, very long processes which contain neuroglia fibres, typical astrocytes, other cells having short, feather-like processes which contain no neuroglia fibres; intermediate forms also occur.

The Distribution of the Spinal Blood Vessels.

The spinal cord is supplied by narrow, long, tortuous arteries derived from the vertebral, intercostal, lumbar, and sacral arteries.[1] Running down the front of the cord, just anterior to the median fissure, is the single anterior spinal artery ; just outside each posterior nerve root is the posterior spinal artery. Encircling the cord on its surface (in the pia mater) is a vascular anastomosis. Fig. 25 shows the vessels entering the cord as seen in a transverse section. The largest are the anterior median arteries, which arise from the anterior spinal artery, run in the anterior median fissure, and then turn to the grey matter of *one* anterior horn.

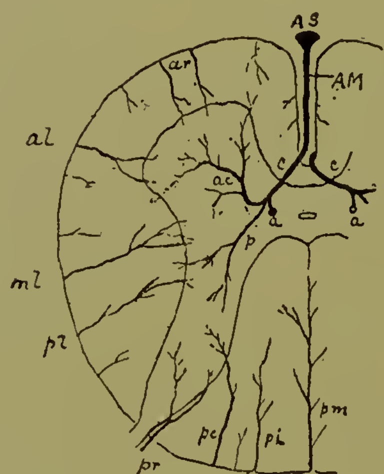

Fig. 25. Arteries of the Spinal Cord.

A S	= Anterior Spinal.	*a l*	= Anterior Lateral.
A M	= Anterior Median.	*m l*	= Median Lateral.
c	= Commissural.	*p l*	= Posterior Lateral.
a	= Anastomotic.	*p r*	= Posterior Root Artery.
a c	= Anterior Central.	*p c*	= Posterior Cornual.
p	= Posterior Central.	*p i*	= Intermediate Septal.
a r	= Anterior Root Arteries.	*p m*	= Posterior Median.

The anterior median arteries and their branches are distributed chiefly to the anterior and central grey matter of the cord, and are often spoken of as the central arteries.

The vessels supplying the cord may, therefore, be divided into two arterial systems :—

1. The *central arteries*—the *anterior median arteries*—entering at the anterior median fissure, and supplying the grey matter, with the exception of the caput and posterior horn.

2. The *peripheral arteries.* These are the small arteries running from the periphery of the cord into the white matter, i.e., towards the centre. The smaller branches end in the white matter, while the larger ones extend to the grey matter.

[1] For more detailed account see writer's small monograph, "On the relation of spinal diseases to the distribution and lesions of the blood vessels of the spinal cord," Lond., 1895, and articles, *Medical Chronicle,* Dec. 1894, and Jan. and Feb. 1895.

But there is no sharp division between these two arterial districts of the cord, since the peripheral part of the grey matter and the adjacent white matter receive arteries from both systems. Hence a transverse section of the cord may be mapped out into three districts (*see* Fig. 26), according to the arterial supply :—

1. The inner part of the grey matter (with the exception of the posterior horn and the caput), which is supplied exclusively by the anterior median artery (area shaded with parallel lines in Fig. 26).

2. The superficial part of the white matter, which is supplied by the peripheral arteries (dotted area in Fig. 26).

3. The peripheral part of the grey matter and the adjacent white matter, which receive arteries from both the peripheral and central systems (area not shaded in Fig. 26). This district, common to the two arterial systems, forms about one-third of the total area of the transverse section of the cord (Kadyi).

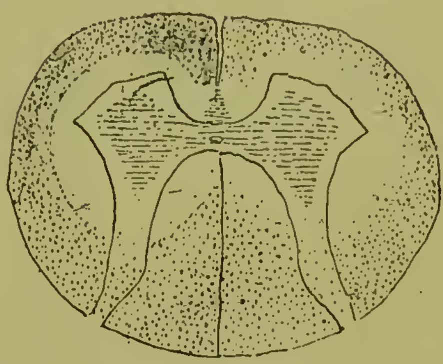

Fig. 26. Section of the Cord showing division into three arterial districts (diagrammatic). Part supplied only by the anterior median and its branches is shaded with parallel lines. Part supplied only by the peripheral arteries is shaded with dots. Part supplied by both systems of arteries is unshaded.

The blood supply of the cord may also be divided into two systems, the anterior and posterior arterial systems ; and the transverse area of the cord may be divided into two districts, according to the origin of the supplying arteries from the anterior or posterior arterial system (*see* Fig. 27).

Fig. 27. Transverse Section of Cord. Area supplied by posterior arterial system shaded with dots. Area supplied by anterior arterial system is not shaded.

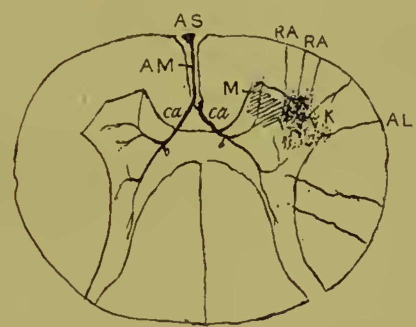

Fig. 28. Section of Cord showing two regions M and K at which changes are most marked in acute anterior poliomyelitis. M (area shaded with parallel lines) supplied by a branch of commissural artery of anterior median. K (shaded by dots) supplied by anterior root arteries (modified after Marie).

In Fig. 28 (modified after Marie), the dotted area represents the part supplied by branches from the *posterior* arterial system ; the unshaded area, that supplied by the *anterior* system.

The grey matter is more richly supplied with blood than the white. The white substance has only a relatively scanty vascular supply, whilst the grey matter possesses a thick network of capillaries. The greater quantity of blood passing into the spinal cord enters the grey matter. According to Kadyi, the arteries penetrating the spinal cord are true terminal arteries (as defined by Cohnheim) and do not anastomose.

The anatomical facts with reference to the blood supply of the cord are of interest as regards the localisation of pathological changes. In infantile paralysis (acute anterior poliomyelitis) the changes are usually inflammatory in nature and are localised often to one anterior horn of grey matter, either around a branch of one commissural artery or around the anterior root arteries as they enter the anterior horn of grey matter

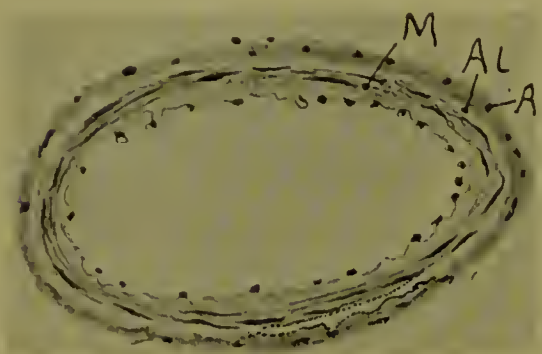

FIG. 29.—Section of a small Spinal Artery stained by logwood and eosin.
A = Adventitial coat.
M = Middle or muscular coat.
On the inner side of this is the intima with elastic lamina. Between the adventitial and muscular coats is the adventitial lymph space, AL.

FIG. 30.—Section of one half of a Spinal Artery, van Gieson's stain.
A = Adventitial coat.
M = Middle or muscular coat.
Between A and M is the adventitial lymph space, AL.

(*see* Fig. 25). The changes in the cord are often found around the branches of one anterior median artery only. This artery usually does not divide, but passes to one horn of grey matter; and hence the inflammatory changes are often found only in one anterior horn. In most cases of acute anterior poliomyelitis of the adult the inflammatory changes are localised to the region of distribution of the anterior median arteries in both anterior horns or to the central area (in Fig. 26), whilst other parts of the cord are unaffected. Often the changes are found both in the cervical and lumbar regions.

In spinal haemorrhage the blood extravasation is very often limited to the grey matter, which is the most vascular part of the cord.

In Friedreich's disease, in ataxic paraplegia, and in many forms of combined postero-lateral sclerosis (or degeneration) of the spinal cord associated with anaemia and various toxic conditions, the pathological changes are chiefly localised to the region supplied by the posterior arterial system (indicated by dots in Fig. 27). Occasionally myelitis

is localised to this region. In "Erb's syphilitic spinal paraplegia" the changes are chiefly in the same region.

The Structure of the Normal Spinal Blood Vessels.

In the arteries of the spinal cord there are three coats as in other parts of the body; (1) the intima or internal coat, (2) the media, (3) the adventitia or external coat. In the intima and the adventitia the

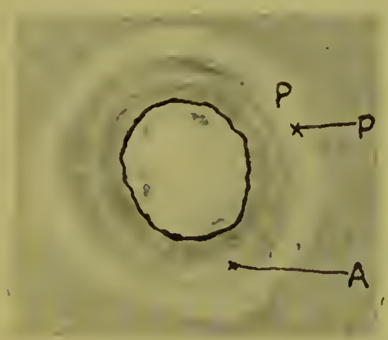

Fig. 31.—Transverse Section of Spinal Artery. Weigert's stain for elastic tissue. Elastic lamina of intima stained deep black. Other structures only very faintly stained.
A = Adventitial lymph space.
P = Perivascular lymph sheath.

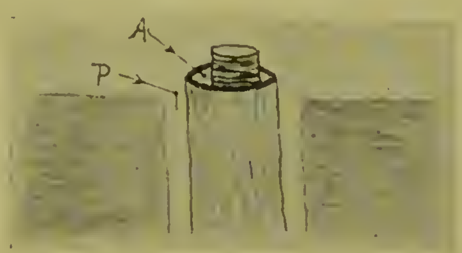

Fig. 32.—Diagram of lymph sheaths of vessels.
P = Perivascular lymph sheath between vessel and cord substance.
A = Adventitial lymph space between adventitia and media.

structures have mainly a longitudinal direction, in the media they are chiefly transverse to the course of the vessel.

In the small arteries (as in those seen in sections of the cord) the internal coat or intima presents an inner layer of elongated endothelial cells arranged longitudinally. External to this layer is a structureless membrane of elastic tissue (without nuclei and cells). The media consists of a layer of involuntary (non-striated) muscle fibres (single in the small arteries). The muscle fibres are transverse to the course of the vessel.

The adventitia consists of connective tissue with longitudinal fibres and a fine network of elastic fibres.

In the medium sized arteries the elastic lamina of the intima is convoluted and the media consists of more than one layer of muscle cells.

In the medium-sized arteries there is a lymphatic space between the media and the adventitia—the intra-vascular or adventitial lymph space. External to the adventitia is the extra-vascular or perivascular lymph space. From this extravascular (perivascular) space the pericellular lymphatic spaces (around the nerve cells) may be injected. The perivascular lymphatic space is bounded externally by a delicate membrane lining the space in the cord substance in which the artery lies (*see* Figs. 29, 30, 31, 32). In the normal condition the perivascular space contains only a few isolated elements—fat globules, pigment granules, lymphoid cells, rarely red corpuscles.

The walls of the vein are thinner than those of the arteries, they

contain less elastic tissue, and the lumen of the vessels is generally greater than that of the corresponding artery. As in the arteries there are three coats. The internal coat consists of endothelial cells. In the muscular coat, the muscle fibres are scanty, and the connective tissue more abundant than in the arteries. The adventitia consists chiefly of fibrous connective tissue with scanty elastic fibres.

The capillaries consist of endothelial cells only, with a closely adherent adventitial sheath.

The *lymphatics* of the spinal cord have been already mentioned. They consist chiefly of sheaths surrounding the blood vessels—the perivascular and adventitial lymph sheaths. The latter communicates externally with the subarachnoid space.

Around the nerve cells are fine spaces, the pericellular spaces. These spaces, and also fine lymph spaces in the nerve tissue, are connected with the perivascular sheaths. Finally the lymph from the perivascular sheath passes into the epi-spinal space—a fine lymph fissure between the glial tissue of the surface of the cord and the pia mater.

Staining of blood vessels in microscopic sections. The external coat can be stained deep red by a weak van Gieson's solution, whilst the internal and middle coats are stained yellow; and thus the lymph sheaths are clearly demonstrated. The elastic coat of the intima is stained deep blue black by Weigert's stain for elastic tissue. Cell infiltration in the walls of the vessels and lymph sheaths is demonstrated well by logwood and eosin.

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SECTION II

GENERAL PATHOLOGICAL HISTOLOGY

Degeneration of Nerve Fibres.

Secondary or Wallerian Degeneration.—When a nerve fibre is divided, the part on the peripheral side of the lesion—the part which is separated from the cell of origin of the fibre—undergoes degeneration; whilst in the part still attached to the cell of origin these degenerative changes do not occur. (The apparent exceptions to the latter statement will be mentioned subsequently.) The same degeneration of nerve fibres occurs when their trophic centres, or cells of origin, are affected by a lesion.

These statements apply to peripheral nerve fibres; but they are also true of nerve fibres in all parts of the nervous system. Hence nerve tracts can be traced in various parts of the central nervous system, by observing the degeneration after experimental or pathological lesions have separated the nerve fibres from their cells of origin. The degenerated fibres are best revealed by Marchi's method of staining, which colours the degenerated nerves black, whilst normal nerves are tinted pale yellow.

After section of a peripheral nerve fibre, *regressive* or degenerative changes occur in the myelin sheath and axis-cylinder, but *progressive* or proliferative changes in the external sheath of Schwann. The latter, of course, are absent in the central nervous system, where the nerve fibres do not possess a sheath of Schwann. In the central nervous system progressive or proliferative changes occur in the neuroglia connective tissue.

After section of a nerve, the axis-cylinder degenerates on the peripheral side of the lesion. The fibrillæ of the axis-cylinder become fused together. The axis-cylinder stains more deeply with carmine; it becomes swollen in some parts, convoluted in others, and is often varicose. Finally it stains faintly and degenerates into granular matter which becomes absorbed later. The peri-fibrillary substance in the axis-cylinder also undergoes granular degeneration.

The myelin sheath becomes broken up into cylindrical or ovoid pieces, and later into small globules.

The degenerated fibres stain deep black in Marchi's method. At first the degenerated myelin forms a black ring around the axis-cylinder;

later it forms irregular black masses and globules, which afterwards degenerate into smaller particles. In the cells of the sheath of Schwann the protoplasm becomes increased in amount, and the nuclei proliferate. These changes reach their maximum during the second week. The degenerated drops of myelin become finally absorbed, and the proliferated cells of Schwann's sheath form spindle-shaped masses, in the course of the degenerated fibres (*see* Fig. 33).

The secondary degenerative changes just described occur more slowly in nerve fibres in the central nervous system than in the peripheral nerves, and in the former situation, for months, the fatty products of degenerated nerve fibres can be seen. Finally they become absorbed, and in the spaces left by degeneration of the nerve fibres scattered granular cells are observed. In course of time the neuroglia increases and this new tissue encroaches on the spaces just mentioned. The degenerated fibres are therefore replaced by a fibrillar structure with a moderate number of nuclei ; still later the new tissue contracts and true sclerosis is produced. Just as in other organs, the degenerated parenchyma is replaced by new connective tissue.

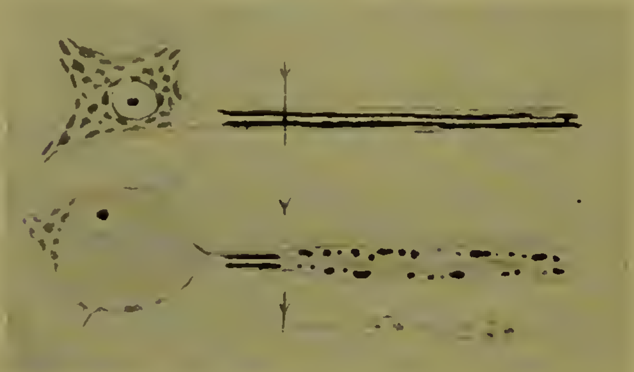

FIG. 33. — Wallerian Degeneration of Nerve Fibre when separated from its cell of origin (diagrammatic). Upper fibre and cell normal. Nissl's granules seen in the cell. Myelin sheath of fibre, deep black. Lower cell and fibre show changes following division of fibre. Myelin is broken up into globules (deep black). Lowest fibre shows final change — collapsed nerve sheath, myelin absorbed. Note spindle-shaped masses of proliferated cells of external sheath (faint black).

SECONDARY DEGENERATION IN THE SPINAL CORD.

When the fibres of the upper motor neurons are destroyed at any point, either in the brain or spinal cord, the portions of the fibres below the lesion are cut off from their cells of origin in the brain, and degeneration occurs in the fibres below the lesion, similar to the Wallerian degeneration in a peripheral nerve. This degeneration is known as descending degeneration ; it can be traced by the method of Weigert when it is marked, by the method of Marchi when it is slight or recent.

Descending Degeneration from Cerebral Lesions.—When the motor cells of the brain cortex, or the fibres coming from these cells (axis-cylinder processes), are destroyed by any lesion, degeneration occurs in the motor tracts below the lesion. In such cases degeneration occurs in the motor part of the internal capsule, in the middle two-fifths of the crusta of the

crus, in the pons between the deep and superficial transverse fibres, and in the anterior pyramid of the medulla. The degeneration down to this level is on the same side as the brain lesion. Below the decussation of motor fibres in the medulla, we find degeneration in the lateral pyramidal tract of the spinal cord on the side *opposite* to the brain lesion, and in the direct or anterior pyramidal tract of Türek on the *same* side as the

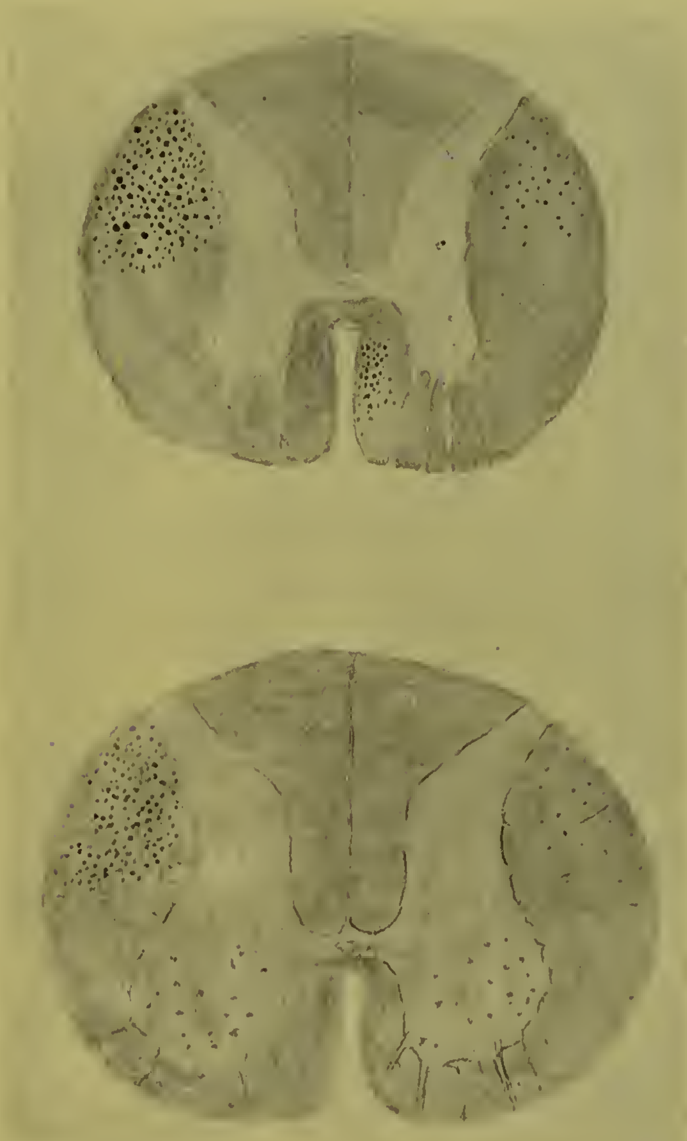

FIG. 34.—Degeneration in Spinal Cord after a Unilateral Lesion of the Cerebrum (tumour of centrum ovale). Marchi's method of staining. Numerous degenerated fibres, deep black, are seen in the lateral pyramidal tract on side opposite to cerebral lesion (left side of figure); a few degenerated fibres (black) are seen in lateral pyramidal tract on same side as lesion, i.e., on right side of figure. Also numerous degenerated fibres are to be seen in anterior pyramidal tract on same side as lesion (right side in figure). Upper figure = dorsal; lower = lumbar region.

brain lesion. The degeneration in the lateral pyramidal tract on the side opposite to the lesion (crossed pryamidal tract) may extend down to the conus medullaris. The degenerated lateral or crossed pyramidal tract is separated by the normal direct cerebellar tract from the

surface of the cord in the lower cervical and dorsal regions ; but in the lumbo-sacral regions it comes to the cord surface.

The degeneration in the direct or anterior pyramidal tract may sometimes be traced down to the lumbar region, or occasionally to the sacral region, though the tract itself, or marked degeneration tehreof, is usually stated to end much higher (*see* Fig. 34). As just described, after a unilateral brain lesion a *marked* degeneration is seen in the lateral or crossed pyramidal tract on the side *opposite* to the lesion ; but, in sections stained according to Marchi's method, very frequently a few degenerated fibres are also found in the lateral pyramidal tract on the *same* side as the brain lesion (*see* Fig. 34.)

The degenerated fibres in the lateral pyramidal tract on the same side as a unilateral brain lesion appear to be almost as numerous below the cervical enlargement as above that level, and Marie thinks this fact supports the view that these uncrossed fibres pass to the lower limb.

Descending Degeneration after Spinal Cord Lesions.—After a transverse lesion of the spinal cord the crossed pyramidal tracts degenerate below the lesion (*see* Fig. 35). There is also degeneration below the lesion in the anterior part of the white matter, close to the anterior median fissure, which extends down to the sacral region. The degenerated fibres in the anterior columns in part belong to the column of Türck or the direct pyramidal tracts ; but most are commissural fibres, connecting different segments of the cord and belong to the anterior marginal descending tract of Löwenthal (or the descending sulco-marginal zone of Marie).

Descending degeneration has also been recorded in other parts of the antero-lateral white matter after a transverse spinal lesion. The following tracts have been described :—

1. Intermedio-lateral fasciculus of Löwenthal—degenerated fibres just in front of, and in part mixed with, the fibres of the lateral pyramidal tract. This tract is between the lateral pyramidal and the lateral cerebellar tract. It does not degenerate in lesions of the brain cortex ; but after al esion at the upper end of the cord—between the cord and the medulla oblongata—degeneration occurs in this tract down to the lumbar region. (Monakow has described a tract of fibres in this region which occupies the same, or almost the same position as the intermedio-lateral tract.) Fibres from the red nucleus probably pass down in this tract.

2. Sulco-marginal anterior descending tract of Löwenthal and Marie, close to the anterior median fissure (already mentioned). (In the same region are fibres of the direct pyramidal tract and also fibres of the ascending sulco-marginal tract.)

3. Close to the anterior nerve root in the cervical region a triangular area of degenerated fibres has been described—Helweg's bundle. It contains fibres which probably come from the cerebral peduncles.

In the posterior columns descending degeneration after a transverse

cord lesion is seen in the following tracts :—(1) The comma-shaped bundle of Schultze. (2) Hoche's bundle. (3) Flechsig's oval field. (4) Gombault and Philippe's triangle.

The tracts 3 and 4 are known as the dorso-medial bundle of Obersteiner, or the septo-marginal tract of Muir and Bruce.

1. Schultze's bundle consists of a comma tract of degeneration

Antero-lateral Columns.

P L = Lateral pyramidal tract.

I L = Intermedio-lateral tract of Löwenthal (Monakow's bundle).

S M D = Sulco-marginal anterior descending tract of Löwenthal and Marie (in this region near the anterior median fissure are the fibres of the direct pyramidal tract and also fibres of the sulco-marginal ascending tract of Marie).

H = Helweg's bundle.

Posterior Columns.

C = Comma-shaped tract of Schultze.

H O = Hoche's bundles.

O = Flechsig's oval field.

G P = Triangle of Gombault and Philippe.

The two tracts last mentioned together form the septo-marginal tract of Muir and Bruce.

FIG. 35.—Descending Degeneration after a Lesion at the upper part of the Spinal Cord (modified after Flatau). Areas of degeneration marked by dots.

First figure = cervical region; second = dorsal; third = lumbar; fourth = sacral.

between Burdach's and Goll's columns. Dorsally it reaches nearly to the periphery, ventrally up to the posterior commissure. Degeneration is found in this tract in the lower cervical, and in the upper and mid-dorsal regions. (The degeneration can be traced for 4 or 5 cm.)

2. Hoche's bundle is a small band of degenerated fibres at the posterior periphery of the posterior columns—close to the posterior septum in the middle and lower dorsal regions (*see* Fig. 35).

3. Flechsig's oval field is a tract of degeneration, seen in the lumbar region, close to the middle of the posterior section of the median longitudinal septum (*see* Fig. 35).

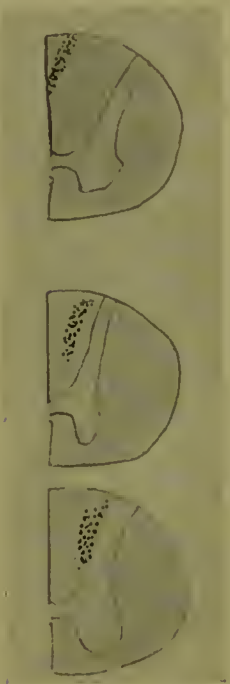

FIG. 36.—Degeneration in the Cord after Section of Posterior Nerve Roots (diagrammatic). Degenerated fibres black. Note degenerated fibres close to posterior grey matter just above lesion (in lowest figure). At a higher level (middle figure) degenerated fibres are nearer middle line. At a still higher level (uppermost figure) they are close to posterior median septum.

4. Gombault and Philippe's triangle is a tract of degeneration in the sacral part of the cord, at the posterior end of the posterior median longitudinal septum (*see* Fig. 35).

After injury to the lower cervical region, or to the upper or mid-dorsal region, descending degeneration is seen in the posterior columns in the region C (the comma-shaped tract). Only in the lower dorsal region are there two tracts of degeneration C and Ho. The comma-shaped tract disappears in the lowest dorsal or lumbar region, and the degeneration collects at the region of the posterior septum and at the dorso-medial angle—Flechsig's oval field and the triangle of Gombault and Philippe.

According to Flatau these four areas of descending degeneration in the posterior columns are parts of one descending tract.

Ascending Degeneration.—Lesion of the posterior nerve roots or of the afferent fibres within the spinal cord is followed by ascending degeneration above the lesion. But in the sacro-lumbar region the dorso-medial or septo-marginal bundle (Gombault-Philippe's triangle and Flechsig's oval field), as well as the ventral field of the posterior columns, do not degenerate when the posterior columns present ascending degeneration. As already described, in man, Goll's column of the cervical region is a continuation of the sacro-lumbar and a portion of the lower dorsal posterior nerve root fibres.

Ascending Degeneration from Lesion of the Posterior Nerve Roots.—In a lesion of a posterior nerve root the degeneration in the cord is first seen on the median side of the posterior horn. At higher levels the degenerated fibres gradually pass to the median and dorsal part of the posterior columns—being gradually separated, more and more, from the posterior horn by healthy fibres.

In a lesion of the lumbo-sacral posterior nerve roots there is first degeneration in the entrance zone of the posterior column close to the posterior horns; at higher levels the degeneration passes gradually toward the middle line, and lies close to the median septum in the cervical

region. In lesion of the posterior roots of the cervical region there is degeneration first in the root entrance zone: the degeneration then gradually passes towards the middle line, and at a higher level reaches the median part of Burdach's column, but Goll's columns are free.

Fig. 37.—Ascending Degeneration after a Spinal Lesion in the Dorsal Region.
G = Goll's column.
B = Burdach's column.
A L G = Antero-lateral ascending tract of Gowers.
C = Lateral cerebellar tract.
S M A = Ascending sulco-marginal tract of Marie.

Ascending Degeneration after Lesions of the Spinal Cord.—Immediately above the lesion both posterior columns are degenerated, with the exception of the parts already mentioned (descending fibres and posterior ventral fields), and of a small zone close to the posterior horn. But very soon the degeneration is limited more and more to the median parts, and only Goll's columns are degenerated. In lesions of the lower part of the cord only the posterior part of Goll's columns are degenerated at the uppermost cervical region.

In the upper dorsal region, the degeneration of Goll's columns often presents a flask-shaped appearance (*see* Figs. 37 and 38A).

In the antero-lateral columns, the direct cerebellar tract and the tract of Gowers present ascending degeneration after a transverse cord lesion above the junction of the dorsal and lumbar regions. In addition to the degeneration described there is, just above the lesion, a diffuse degeneration of the whole antero-lateral columns: in the anterior column it is most marked just around the anterior horn; at a higher level the degeneration passes to the periphery, and the part around the anterior horn becomes free (*see* Fig. 37). Adjacent to the anterior median fissure this degeneration forms the sulco-marginal ascending tract of Marie.

In transverse lesions of the cord below the junction of the dorsal and lumbar

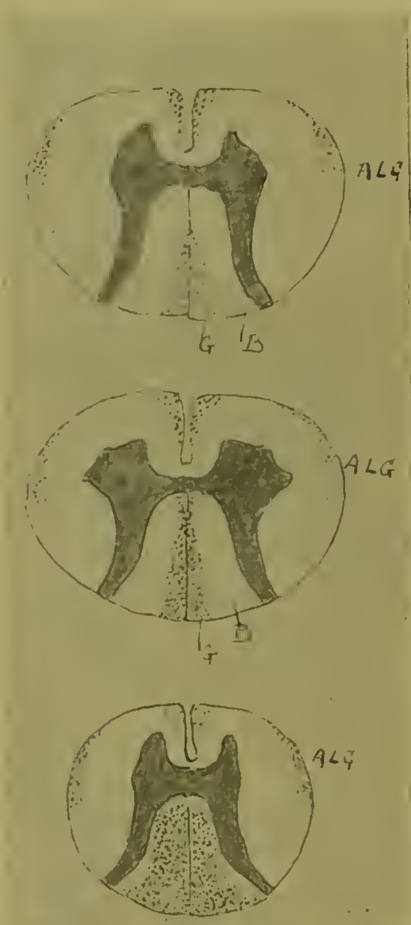

Fig. 38.—Ascending Degeneration after a Lesion in the Middle Lumbar Region of the Cord.
Lowest figure = the lower dorsal region just above lesion. Middle figure = the lowest cervical region. Uppermost figure = the upper cervical region. Note that direct cerebellar tracts are *not* degenerated. Lettering same as in preceding figure.

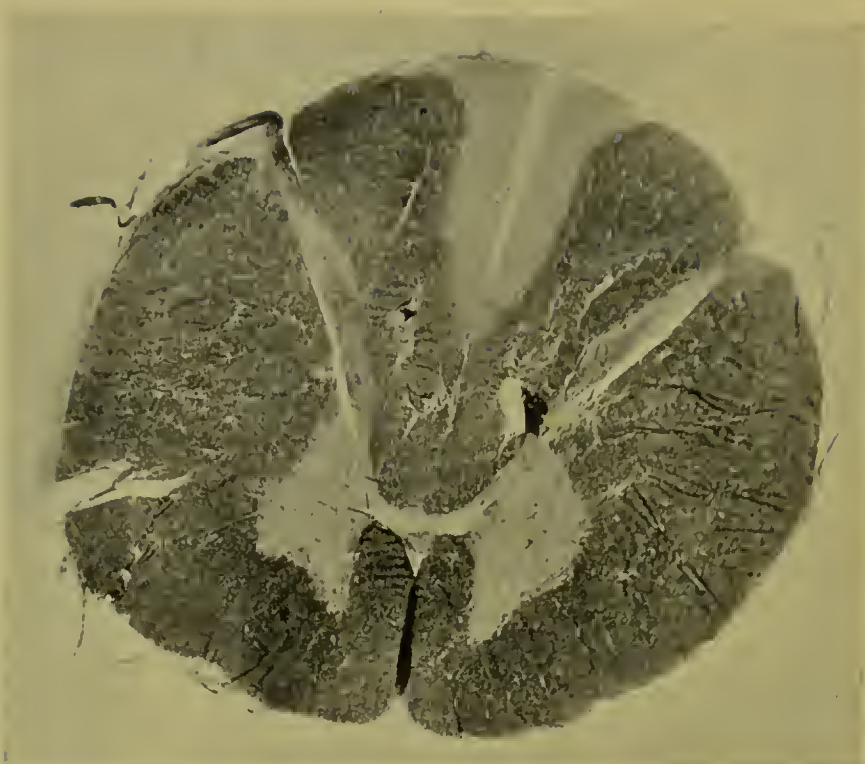

FIG. 38A.—Ascending Degeneration in Posterior part of Goll's Columns. Section of upper cervical region of Cord. Weigert's stain. Degenerated tracts pale. Note anterior part of Goll's columns are unaffected. The primary lesion was a compression myelitis at the lower lumbar region of the Cord.

FIG. 38B.—Ascending Degeneration in Goll's columns. The photomicrograph is taken from the same section as the preceding figure, but is more highly magnified. To the left of the photograph is the pale degenerated tract with but a few scattered (black) nerve fibres. To the right is the normal posterior column presenting the nerve fibres (black) in transverse section (Weigert's stain). (The clear space near the left margin of the photograph is a fissure in the section just in the position of the posterior median septum.)

regions (i.e., in the lumbar enlargement) the direct cerebellar tract does not degenerate since this tract only arises at the level mentioned (*see* Fig. 38); but the antero-lateral ascending tract of Gowers degenerates in these lesions.

If the lesion be in the cauda equina or lumbo-sacral posterior nerve roots, the antero-lateral tract of Gowers is unaffected by degeneration, but the posterior columns degenerate in the usual manner.

Degeneration of the Spinal Cord following Lesions of the Cerebellum.—Marchi has found that removal of one half of the cerebellum in the dog is followed by degeneration in the peripheral part of the antero-lateral

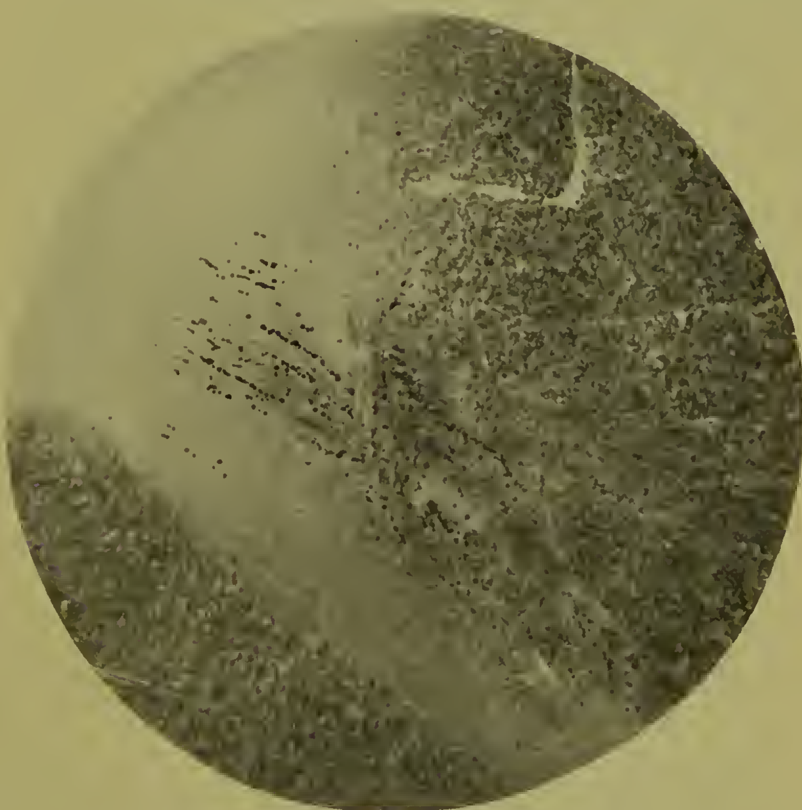

FIG. 39.—Section of Cervical Cord from a case of Cerebellar Tumour. Showing the posterior horn of grey matter at the posterior part of the Cord (pale portion just to the left of the middle line). On the median side (right side of the figure) of the posterior horn and near the centre of the figure are the fibres of the posterior root (black dots), showing degeneration directly they have entered the cord.

column. It is most marked at the upper part of the cord, but can be traced down to the lower part. This degeneration has, however, been much disputed.

Spinal Changes associated with Intra-cranial Tumours (Cerebral and Cerebellar), and other Conditions.—In addition to the ordinary descending degeneration, produced by tumour growths involving the motor tracts or areas of the brain, changes have also been described, in some cases, in the posterior columns.

Batten and Collier have carefully studied these changes. They found degeneration of the posterior columns of the spinal cord in about

65 per cent. of the cases of intra-cranial tumour examined. The changes were independent of the situation of the tumour, and were observed both in cerebral and cerebellar growths. They were best seen in the

Fig. 40.—Degeneration in Burdach's Columns of the Cord, Cervical Region; following degeneration of the extra-medullary fibres of the posterior nerve roots at the upper part of the cord, in a case of cerebellar tumour. Marchi's stain (diagrammatic). The preceding figure was from the same case. The intra-medullary fibres of the posterior roots at the lower part of the cord were not affected.

cervical region and affected the postero-external column more than the postero-internal column. The degeneration is of root origin, and arises from the point where the posterior root enters the cord. The posterior roots outside the cord are always less affected than the posterior

Fig. 41.—Microphotograph of Cervical Region of Spinal Cord from a case of Cerebral Tumour in the third Ventricle. Marchi's stain. Note the degenerated fibres in the posterior columns (black dots). They are most numerous close to the posterior horns of the grey matter. Degeneration commenced in the fibres of posterior roots directly they had passed to the inner side of the spinal pia mater.

columns, and they may present no degeneration. Degeneration may also occur in the direct cerebellar tract.

The loss of knee jerks in some cases of intra-cranial tumour may be due to these changes in the posterior root fibres. I have found a

FIG. 42.—Microphotograph of Lumbar Region of the Spinal Cord in the same case as the preceding. The same description applies.

similar degeneration of the intra-medullary fibres of the posterior nerve roots, commencing directly these roots have passed through the pia mater, in some severe cases of diabetes mellitus. Also, the same form of degeneration has been found in general paralysis of the insane, and in several other conditions (*see* p. 377).

Law of Eccentric Position of Long Tracts in the Spinal Cord.—The observations of Schiefferdecker, Flatau, and others, have shown that in man and the higher mammals the short ascending and descending fibres in the cord are situated close to the grey matter, and that the long fibres tend to pass towards the periphery of the cord. In the posterior columns the fibres running the longest course gradually pass to the posterior and median part of the posterior columns (in Goll's columns). The tendency of the long fibres to assume this position has been described by Flatau as the "*law of the eccentric position of the long tracts in the spinal cord.*"

This same tendency is shown clearly in the spinal degeneration

following experimental compression of the abdominal aorta in animals. Sarbó found, in the lumbar region of the cord, a diffuse degeneration in the white matter just around the anterior and median grey substance; but in the dorsal and cervical regions the degeneration had passed to the periphery of the cord.

Primary Degeneration of Nerve Fibres.

Certain poisons have a direct localized action on nerve fibres in different regions of the nervous system, and cause degeneration which may be described as primary degeneration. The nerve fibres are not separated from their cells of origin, and the cells are not destroyed in such cases. Other poisons circulating in the blood act also on nerve cells, or on both cells and fibres. When the cell is affected secondary degeneration of the nerve fibre may follow.

If the injurious action of the poison is not too great, a pure diffuse primary change in the nerve may follow. But if the toxic action be severe, the axis-cylinder degenerates, and a secondary or Wallerian degeneration will then follow on the peripheral side of the degenerated region. The more diffuse and slight the toxic action, the more pure the form of primary degeneration.

In disseminated sclerosis, at first only the medullary sheath of the fibres is affected; naked axis-cylinders are seen in the diseased patches; and in such cases secondary degeneration does not usually occur.

In Wallerian, or secondary degeneration, all fibres degenerate at once on the peripheral side of the sudden lesion separating the fibres from their cells of origin. In primary degeneration the process is gradual; fibre after fibre gradually degenerates; and hence the fibres are not all affected to the same extent.

When the toxic or injurious action on the nervous system is very slow, slight, and diffuse, the most characteristic form of primary degeneration occurs. There is then a slow and progressive atrophy of the myelin sheath, which extends regularly from the periphery of the fibre towards the cell of origin. Changes in the axis-cylinder only occur at a late stage of the disease.

In some cases Marchi's method of staining does not reveal these primary changes; but staining with nigrosin or safranin shows the lesion well. The myelin sheath becomes small, the margins of the axis-cylinder become indefinite. The axis-cylinders present varicosities and constrictions. Primary changes are best marked in the pyramidal tracts and posterior columns.

In other cases, at the early stage the degeneration is revealed by Marchi's method, later by Weigert's stain. The axis-cylinder may become swollen and the myelin sheath transformed into granular globules. In rapid degeneration granular globules are abundant, in chronic processes few or absent. At a late period spaces are found in the neuroglia from which the nerve fibres have disappeared, and, in addition, scattered

swollen fibres are seen with dilated myelin sheaths which stain feebly. In time the neuroglia increases and a true sclerosis is produced.

Toxic substances may have an elective action on nerve fibres in various parts of the nervous system. Thus the nerve fibres of the posterior column of the cord, chiefly the intra-medullary fibres of the posterior roots, degenerate in tabes, ergotism, some severe cases of diabetes mellitus, some cases of intra-cranial tumour, etc. (*see* p. 37). In multiple neuritis from alcoholism, diphtheria, lead, and other causes, in leprosy and beri-beri, changes may be sometimes found in the posterior columns or posterior nerve roots in addition to the neuritic changes in the peripheral nerves.

In severe anæmia, in cancer of various organs, pellagra, Addison's disease, leucocythæmia, and in several other affections degeneration of nerve fibres has been observed in the posterior columns or in both posterior and lateral columns (*see* pp. 362 to 377).

The intensity of these changes varies. In some cases there is total degeneration of the nerve fibres and increase of the neuroglia ; in other cases simple thickening of the axis-cylinder and dilatation of the medullary sheath.

In several tract diseases, as in tabes and amyotrophic lateral sclerosis, there is primary degeneration of nerve fibres.

Indirect or Secondary Changes in Nerve Cells.[1]

Motor Neuron.—After the *division of a peripheral motor* nerve not only do the nerve fibres undergo degeneration on the peripheral side of the lesion (Wallerian degeneration), but changes may also be detected in the motor nerve cells in the spinal cord from which the nerve fibres take their origin. To these changes the names of chromatolysis, tigrolysis, retrograde degeneration, réaction à distance, and degeneratio axonalis have been given.

The changes in the motor nerve cell, after section of a motor peripheral nerve, may be divided into two stages :—

1. The stage of solution of the chromophile substance. The Nissl's granules (or chromophile substance) become gradually dissolved. The disappearance of these granules begins in the centre of the cell and gradually extends to its periphery. The cell stains more or less uniformly ; but often at the periphery a few Nissl's granules may be still seen (*see* Plate II, Fig. 2). The whole cell becomes enlarged ; the outlines, instead of being concave, become convex, and the cell has a swollen or distended appearance. The nucleus, instead of occupying its normal central position, passes to the periphery of the cell. These changes may be seen 40 hours after section of a motor nerve, and they can be detected for 15 to 20 days. Some cells may undergo atrophy and disappear entirely.

2. When the cell does not completely degenerate the stage of repair

[1] This description of changes in nerve cells is based chiefly on the writings of Van Gehuchten.

DESCRIPTION OF PLATE II.

1. Normal nerve cells of anterior horn of grey matter of spinal cord. Toluidin-blue stain. Note deeply stained chromophile substance (Nissl's granules). Axis-cylinder process of cells to the left is free from granules.

2. Nerve cells showing central chromatolysis (tigrolysis). Note swollen appearance of cells; absences of chromophile substance, except at periphery of cell; also nucleus displaced to the side of the cell. Toluidin-blue stain.

3. Nerve cells showing peripheral chromatolysis. Note absence of chromophile substance at periphery of cells. Toluidin-blue stain.

4. Nerve cells showing finely granular condition of chromophile substance. Toluidin-blue stain.

5. Nerve cell showing pyknosis.

6. Nerve cells with pigment granules. In the illustration the pigment granules appear deep black; in the specimen they were yellow. Absence of chromophile substance in region of pigment. Toluidin-blue stain.

7. Nerve cells (Marchi's stain), containing fine pigment granules, stained black.

8. Calcareous degeneration of nerve cells.

9. Simple atrophy of nerve cells (case of progressive muscular atrophy); one cell (to the right) contains much pigment. Aniline blue black stain.

[Figs. 1, 2, 3, 4, 6, 7, and 9 are from my own specimens. Fig. 5 is modified after Schmaus; Fig. 8 is modified after Obersteiner.]

PLATE II.

occurs; the chromophile granules gradually return, and become thicker and more numerous than in the normal condition ; the cell diminishes in size, and gradually returns to the normal volume ; the nucleus returns from the periphery of the cell to its normal position in the centre. Apart from the change in position, the nucleus remains unaltered during the whole period.

The changes just described may be seen in the hypoglossal nucleus of animals after division of the hypoglossal nerve. In man the course of the changes is not quite the same as in animals. The cells in the anterior horns of the human spinal cord present the changes of chromatolysis when examined three to seven months after amputation of a limb. In man the stage of solution of the chromophile granules is not followed by a stage of repair. The changes proceed to atrophy of the nerve cells. In man section of a peripheral motor spinal nerve is always followed by very distinct changes in the motor ganglion cells of the cord. In experiments on animals the changes of chromatolysis always occur after section of a motor cranial nerve ; but after division of a motor spinal nerve chromatolysis in the motor nerve cells is sometimes absent ; in other cases, however, it has been detected.

Sensory Neuron.—After section of a peripheral sensory nerve in animals (i.e. section of the peripheral processes of spinal posterior root ganglion cells) chromatolysis, similar to that just described, is seen in the nerve cells of the ganglion of the posterior nerve root. Usually recovery occurs, but sometimes the changes proceed to destruction of the cell. But when the posterior nerve root (sensory root) is divided between its ganglion and the surface of the cord, no cell changes in the ganglion can be detected. Further, in man, disease of the nerve fibres of the posterior columns is not associated with changes in the cells of the ganglia of the posterior roots.

Central Neurons.—In the dog, division of the fibres of a *central motor neuron* (i.e. fibres coming from the nerve cells of the motor cortex) is followed by chromatolysis of the large pyramidal cells of the motor cortex of the brain. These changes are followed by complete destruction of the cell. The same reaction has been observed in the cells of the motor brain cortex in man, after destruction of the motor fibres in the internal capsule, and complete atrophy of large pyramidal cells has been noted. After division of the axis-cylinder process (nerve fibre) of a central motor neuron, the chromatolysis in the nerve cells is followed by cell degeneration ; whilst the changes in the motor cells, after division of the axis-cylinder process of a motor peripheral neuron, are followed by repair or regeneration in animals.

A comparison of the peripheral and central neurons shows, that in animals the peripheral motor neurons of the spinal cord have the greatest resisting power. In animals, section of the axis-cylinder process (axon) of a peripheral motor neuron (motor spinal nerve) is not always followed by chromatolysis in the nerve cell ; section of the axon of a peripheral cerebral motor neuron (cranial nerve) produces chroma-

tolysis in the nerve cell, and this is followed by repair; the motor cortical neuron degenerates most easily, since section of its axis-cylinder process is followed by complete degeneration of the cell.

With regard to the changes in the *central sensory neuron*, after section of the axon, less definite statements can be made. One point, however, has been clearly determined—compression or section of the fibres of the lateral cerebellar tract is followed by chromatolysis of the cells in the column of Clarke. A general characteristic of the changes in the central neurones (both motor and sensory) is atrophy of the nerve cells after section of the axons.

W. B. Warrington (of Liverpool) has shown that section of sensory spinal nerve roots is followed by chromatolysis in the posterior lateral group of nerve cells of the anterior (motor) horn of grey matter. (*See Journal of Physiology*, vols. xxiii. and xxiv. See also Brauenig, *Arch. f. Anat. u Phys.*, 1903, p. 251. Abstract in *Review of Neurology and Psychiatry*, Sept., 1903.)

From a careful study of the chromatolysis and other changes in nerve cells of the spinal cord after the amputation of limbs or portion of a limb in the human subject, the motor functions of the ganglion cells of the anterior horns have been mapped out. (A. Bruce, *Scottish Med. and Surg. Journ.*, Dec. 1901.)

Primary Changes in Nerve Cells.

The changes just described are secondary. But *primary* changes are also found in the nerve cells in various pathological conditions. The following are the chief primary changes in the nerve cells of the spinal cord :—

1. Fine granular transformation of the tigroid or chromophile substance—producing a dusty appearance.

2. Swelling and increase of the chromophile substance—a rare condition.

3. Fusion of the chromophile granules into irregular masses, which collect at the centre of the cell, whilst the periphery of the cell is often free from granules.

4. A common change is solution of the chromophile substance, so that the cell appears pale—chromatolysis or tigrolysis. This chromatolysis may begin at the periphery of the cell and pass to the centre, or it may occur in a diffuse manner, or it may begin at the centre and pass to the periphery. In some cases the chromatolysis is partial, occurring at the periphery of the cell only, or at the centre only, whilst the nucleus remains in its normal position in the centre of the cell. In other cases the chromatolysis affects the whole of the cell, which stains feebly and uniformly, and the nucleus is altered in position just as in the secondary or indirect changes after section of nerve fibres.

5. A more profound change consists of thickening of the chromophile substance, with diminution of the size of the cell. Both the Nissl's bodies and the substance between them stain well; the two are blended

together, and the whole cell stains more deeply. The nucleus also stains more deeply; it becomes smaller, and finally melts away into the body of the cell. The processes of the cell also stain deeply. To this condition the name of *pyknosis* is given. As the changes progress the cell becomes smaller and its contour sharper. Finally the cell is transformed into a small irregular mass without processes, and the staining becomes gradually paler.

6. Swelling of the cell. The outline becomes rounded and distended; between the cell processes it is convex instead of concave; the processes also become thicker.

7. Vacuoles are sometimes seen in the cell protoplasm. They are usually at the periphery. If numerous the cell may present a honeycombed appearance.

8. Molecular or granular degeneration. The cell protoplasm is studded with irregular or roundish particles, causing a dusty turbidity or a homogeneous swelling of the cell. Fatty degeneration may finally occur, but the fat granules are often dissolved by the alcohol with which the sections are treated.

9. Pigmentation of nerve cells is a common change. At one part of the cell yellowish pigment is seen in the cell protoplasm. The chromophile substance is absent at this part. Pigmentation of cells is a common senile change, but it may occur in pathological conditions which are not senile. Both pigment and fat in nerve cells stain black with osmic acid. In sections stained according to Marchi's method pigment granules in the nerve cells also stain black. If specimens stained with osmic acid are treated with warm xylol, fat granules are dissolved, but pigment granules remain.

10. In calcareous degeneration the cells are infiltrated with lime salts, and the addition of a little dilute acid to the section causes small bubbles of gas to be given off.

11. Finally the condition of simple atrophy may be mentioned. The cells become smaller and lose their processes; some cells stain deeply, others faintly.

Reaction Appearances.—In many chronic pathological conditions nerve cells simply atrophy, and no change takes place in the surrounding elements. In other cases, when the death of the cell is more rapid, as in many toxic and infectious conditions, a reaction occurs in the surrounding tissue; new cells form around the diseased nerve cells, and penetrate the cell substance of the latter. To the new cells the name of "neuronophagen" is given, and they are believed to be of neuroglia origin. These changes may be well seen in the sympathetic ganglia.

The **nuclei** of nerves cells may present various changes. When these are marked the pathological lesion is severe. Disappearance of the nucleus indicates death of the cell. The position of the nucleus is often altered, and this change accompanies swelling of the cell, as already mentioned; it is displaced to one side of the cell in secondary changes

after division of nerve fibres; sometimes its position is altered in primary changes. The nucleus is sometimes swollen; it may lose its power of staining with various dyes (karyolysis), or the arrangement of the nuclear chromatin may be altered. In many pathological conditions the nucleus stains deeply and in a homogenous manner.

* * * * *

Causes of Chromatolysis.—Secondary chromatolysis is due to a lesion of nerve fibres connected with the cells. Primary chromatolysis is caused by the action of high temperature, or of toxic substances in the system; also it is found in many diseased conditions. Thus primary chromatolysis has been caused by the toxic action of phosphorus, arsenic, chlorate of potash, alcohol, strychnine; it has been found in various diseases and diseased conditions, as in uræmia, diabetes, tetanus, diphtheria, general paralysis of the insane, alcoholism, senile dementia, tuberculosis, cancer, heart failure and pernicious anæmia. In spinal softening, acute myelitis and ascending paralysis, chromatolysis has also been observed. The frequency with which it has been found in so many diseases diminishes its importance to the clinician.

So-called "Retrograde Degeneration."

According to Waller's law, as already mentioned, after the division of a nerve (1) the fibres on the peripheral side of the lesion degenerate, whilst (2) the fibres on the central side of the lesion—those still connected with the trophic centre—do not degenerate. Certain apparent exceptions to the second part of this law have been recorded, and to these unusual changes the name of retrograde degeneration has been given. Lugaro gives the following instances of this so-called retrograde degeneration :—

1. Krause and Friedlander have observed, after division of a mixed nerve, that a *small* number of degenerated fibres may be detected, by Marchi's method of staining, in the central part of the divided nerve. These changes occur rapidly.
2. After amputation of limbs (in man or animals) changes of a degenerative or atrophic nature occur in the nerves of the remaining portion or stump of the limb. These are of the nature of a diffuse and slow atrophy. The nerve cells of the anterior horn degenerate also, and often the change is localised to one group of cells.
3. Gudden has shown, that in new-born animals division of a nerve fibre is followed by marked atrophy of the central portion, and degeneration of the cells of origin, as well as by Wallerian degeneration of the fibres on the peripheral side of the lesion. ("Gudden's Method" of experimental study of the nervous system). These changes occur rapidly.
4. Occasionally degeneration has been observed in nerve tracts, extending in a direction the opposite of that in which the fibres conduct nervous impulses. Thus, after disease of the nerve cells of the anterior horn, ascending degeneration has been occasionally observed in the pyramidal tracts. This is an exceptional form of degeneration, the changes are very chronic and consist in simple atrophy of the myelin sheath of the nerve fibres.

As Lugaro points out, the facts just recorded do not, however, disprove Waller's law of degeneration.

The changes in the first group are apparently the result of an ascending neuritis, due to interstitial inflammation of the nerve creeping along the course of the lymphatics. In the second and third group of cases death of the trophic nerve cells (those of the anterior horns of grey matter) is probably the cause of the nerve fibre degeneration. Following division of the nerve chromatolysis has occurred in the nerve cells, and this has led to atrophy and death of the cells. True Wallerian degeneration has then occurred in the fibres arising from these cells.

The changes in the fourth group are probably due to the primary action of a toxic substance circulating in the blood.

Changes in the Spinal Cord and Nerves after Amputation of a Limb.

After the amputation of a limb, or a portion thereof, changes have been found in the spinal cord—in the cervical region when the arm has been amputated, in the lumbar region in amputation of the leg. Occasionally the half of the cord on the side of the lesion has been smaller than the other half, the change affecting both the anterior and posterior white and grey matter; in other cases only the anterior horns and antero-lateral columns have been affected; whilst in a third group of cases, only the nerve cells in the anterior horns have been degenerated, and often only a special group of these nerve cells has been affected.

After some months, the motor nerves of the limb above the seat of amputation have been found atrophied. Microscopically, besides normal fibres, thin fibres with very small, badly stained, medullary sheaths have been observed.

Histologically, in most cases after amputation, the changes in the nerve fibres are of an atrophic nature, and consist of thinness of the separate fibres and their sheaths; but fatty degeneration of the myelin sheath with subsequent sclerosis is usually absent. Thus the change is usually a pure quantitative one: whilst the Wallerian degeneration is a qualitative change. This statement holds good for most cases, but in a few true degeneration of nerve fibres has been observed.

After experimental amputations in animals, however, distinct degeneration of the nerve fibres, resembling Wallerian degeneration has been observed. Also, after some months, there is atrophy of the posterior horns and posterior columns, of Clarke's column, of the cells of the anterior horns, of the sensory nerve fibres immediately before their entrance into the spinal ganglia, and of fibres within the posterior ganglia. In some cases degenerative changes in the peripheral nerve fibres have been observed; in other cases simple atrophy of fibres. The latter is probably only a more chronic and less intense process than the Wallerian degeneration. Both are probably due to the changes in the nerve cells of origin which follow sections of the nerve. These changes—chromatolysis—in time terminate in recovery in some cells; but in others they pass on to death of the cells (*see* p. 43).

After section or transverse lesion of a nerve, when the fibres unite, probably the nerve cells return to the normal condition; when no union occurs the cells degenerate and degeneration of the central portions of the divided fibres follows. The so-called Retrograde Degeneration does not always occur; the degeneration does not affect all the fibres of the central portion (whereas all fibres of peripheral portion of a divided nerve show the Wallerian degeneration); moreover, it is limited to a number of fibres, and histologically there is usually a less degree of degeneration than in true Wallerian degeneration.

Tertiary or Transneural Degeneration.—After amputation of a limb there is atrophy of the posterior root; also atrophy of the cells

of Clarke's column may occur. Atrophy of the cells of the anterior horns has been observed after lesion of the posterior roots. To these changes the term of tertiary degeneration has been applied.

Pathological Changes in the Neuroglia.

Disease of the nerve elements often causes increased activity of the neuroglia cells. There is a return of the partially, or totally, lost energy of the neuroglia cells (*see* p. 22). The nucleus proliferates, the protoplasm becomes increased, the cell becomes separated from the old neuroglia fibres and produces new fibres.

In some cases when the nerve elements are affected first, as in true secondary degeneration of nerve tracts, the neuroglia changes are simply secondary.

In other cases it is not possible to say, to what extent the neuroglia changes are dependent on the degeneration of nerve elements, and to what extent they are due directly to the primary pathological conditions.

In cases of extensive and severe lesions both nerve elements and neuroglia degenerate. In chronic pathological conditions, or in changes of slight intensity, the increased activity of the neuroglia cells acts injuriously on the nerve element; the neuroglia cells enlarge, their nuclei proliferate, and the cells separate from the neuroglia fibres.

By some pathologists compound granular cells are regarded as neuroglia cells separated from the fibres.

In secondary degeneration of nerve fibres and cells the increase of the nuclei of the neuroglia is slight: in chronic toxic conditions the neuroglia proliferation is greater, but it is greatest after true inflammatory processes.

In some forms of chronic sclerosis there is simply an increase of the neuroglia fibres, and very little increase of the glial cells. But in other more active conditions there is marked increase of the cells.

Very large neuroglia cells, with processes—monster cells—are sometimes seen when the cells are in a state of great activity.

Amyloid bodies—round or oval structures with dark margins—are probably produced from neuroglia cells.

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SECTION III

FUNCTIONS OF THE SPINAL CORD

Briefly stated the following are the chief functions of the spinal cord :—

1. It conducts motor (or efferent) impulses from the brain, and sensory (or afferent) impulses to the brain.

2. It contains "centres" for reflex actions.

3. According to many physiologists, it contains the reflex centres for the bladder, rectum, and the viscera, or part of these centres.

4. It contains also vaso-motor and trophic centres.

As regards the conduction of the motor and sensory impulse, it was shown by Sir Charles Bell, long ago, that the anterior nerve root contained motor fibres and the posterior sensory fibres.

Conduction of Impulses from the Brain.—Motor impulses are conducted from the brain downwards in the lateral and anterior pyramidal tracts. The impulses finally pass along fibres which proceed from these tracts to the anterior horns of grey matter, and there stimulate the anterior ganglion cells. From these cells impulses pass along the anterior nerve roots and motor nerves to the muscles (*see* Fig. 17).

According to some neurologists, the fibres from the pyramidal tracts do not actually enter the anterior horn, but pass into the region of the posterior grey matter, and a short intermediate or connecting neuron transmits the impulse to the anterior horn (*see* Fig. 18).

A few motor fibres which do not decussate at the medulla, pass into the lateral pyramidal tract and probably supply motor impulses to the leg on the same side as the cerebral cortex from which the motor fibres arise (*see* Fig. 16).

Lesion of the lateral pyramidal tracts in the spinal cord causes : (1) paresis or paralysis of the limbs ; (2) rigidity (spastic condition) of the affected muscles ; (3) increase of the reflexes, ankle clonus, and the extensor form of plantar reflex. (4) There is no muscular atrophy (except slight wasting from disuse). (5) There is no reaction of degeneration on electrical examination.

In addition to the conduction in the pyramidal tracts impulses pass, in animals at least, from the brain down into the spinal cord in Monakow's bundle of fibres (*see* Fig. 35).

The anterior ground bundle of nerve fibres, the lateral limiting layer and the posterior ventral field probably contain commissural or endo-

genous fibres, i.e. fibres which connect or link together the various segments of the grey matter of different levels.

The ganglion cells in the anterior horns of grey matter are the trophic centres for the muscles. From these ganglion cells arise the fibres of the anterior nerve roots. These nerve cells are interposed in the course of the fibres which form the paths for the impulse producing reflex movements, i.e. they form, therefore, a part of the arc of fibres which must be completed in order that certain reflex movements shall occur.

Lesion of the ganglion cells of the anterior horns causes the following symptoms :—

1. Paralysis of muscles supplied from these cells, with flaccid condition of the muscles.

2. Atrophy of the paralysed muscles.

3. Loss of the reflexes at the level of the lesion. (The plantar reflex, if present, is of the normal flexor type.)

4. Certain electrical changes, known as the reaction of degeneration, in the paralysed muscles.

Definite functions, as regards movements and as regards the motor supply of the muscles, have been localised to various spinal nerve roots and segments of the cord. These motor functions have been determined : (1) Through experiments on animals—various nerve roots being divided and the extent of the paralysis observed ; (2) by clinical and pathological observations in lesions of nerve roots or segments of the cord at different levels ; and (3) by noting the change in the nerve cells of the anterior horns after amputation of sections of a limb, and in congenital absence of a limb or segment thereof.

Not only have definite motor functions been localised to various segments of the cord in the vertical direction, but by some observers definite functions have also been attributed to various groups of nerve cells (*see* p. 107) through the results of the last mentioned method of investigation. Though the exact localisation of the functions is disputed by certain physiologists, the general results of spinal localisation, both in its vertical extent and as regards the groups of nerve cells, are of much practical value.

Some neurologists believe that the descending fibres in the posterior columns forming the oval bundle of Flechsig, or a portion of these fibres, convey motor impulses to the bladder and rectum.

Conduction of Impulses to the Brain.—All afferent impulses (producing the sensations of touch, pain, and temperature and afferent impulses from the muscles, joints and bones), pass to the cord from the peripheral sensory nerves through the posterior nerve roots. They are then conducted to the brain or to other neurons within the cord.

As already described some of these impulses entering by the posterior roots pass through the " reflex collaterals " to the nerve cells of the anterior horn ; others pass to Clarke's column and then to the direct cerebellar tract and upwards to the cerebellum. Others pass upwards

in the posterior columns to the medulla, and thence chiefly to the opposite cerebral hemisphere in the fillet, a minority to the cerebellum. It is believed by many that the grey substance of the posterior horns conducts sensory impulses, and that some of the impulses from the posterior roots pass to nerve cells in the posterior horns, and from these by fibres running through the anterior commissure to the antero-lateral column of the opposite side of the cord, and then pass upwards, in this region, to the brain.

The conduction paths for the various forms of sensation are disputed; for many reasons it is difficult to arrive at a definite conclusion, and only a brief summary of the different views can be here given.

In the first place, it is a curious fact, that, in lesions of the spinal cord, motor fibres appear to be so much more readily injured than sensory. Very frequently we find marked motor paralysis, whilst sensation is not impaired or only slightly impaired.

Again we not infrequently find anæsthesia to one kind of sensation, whilst sensation to other kinds of the impressions is unimpaired. Thus loss of sensation to pain and temperature may occur, whilst sensation to tactile impression is unimpaired. This is often the case in syringomyelia. This fact would appear to indicate, that the path for the conduction of pain and temperature is not the same as that for the conduction of tactile impressions.

When there is loss of sensation to pain there is generally also thermo-anæsthesia, and hence it has been supposed, that both the tracts for the conduction of painful impressions and for the sense of temperature run close together.

It has long been taught, that the sensory paths cross soon after entering the cord to the opposite side, and then ascend to the brain. This view was based on—(1) the experiments of Brown-Séquard, in which, when one half of the cord was divided, anæsthesia was produced on the opposite side of the body; and (2) on the results of clinical and pathological observations, which show that a lesion of one-half of the cord produces paralysis on the same side and anæsthesia on the opposite side.

The crossing of sensory fibres has been much disputed. The results of many experiments on animals have been published, by various physiologists, which are held to favour the view that most of the afferent fibres do not cross in the spinal cord. But W. A. Turner and others have published the results of experiments, and collected numerous pathological records, which confirm the old view of the decussation of sensory fibres.

Whatever may be the exact course of the sensory fibres in the cord, it may be accepted that disease of one half of the cord gives rise to paralysis on the same side, and anæsthesia, as regards pain and temperature at least, on the opposite side.

The following is a statement of the views most commonly accepted with respect to the course of sensory impulses :—

The afferent impulses related to the muscular sense probably pass upwards to the brain in the posterior columns of the same side as the muscles in which they originated. The impulses from the legs are probably conducted in Goll's columns: the impulses from the arms in Burdach's columns of the cervical region. Degeneration of the posterior columns in spinal diseases causes loss of muscular sense, but little affection of cutaneous sensibility. Tactile sensation (from the skin) is believed by some observers to be conducted upwards in the posterior columns of the same side as the part of skin touched. But many believe that soon after entering the cord the impulses are conducted to the opposite side of the spinal cord, by means of a secondary neuron (cell and fibre). Others believe that tactile impressions are conducted

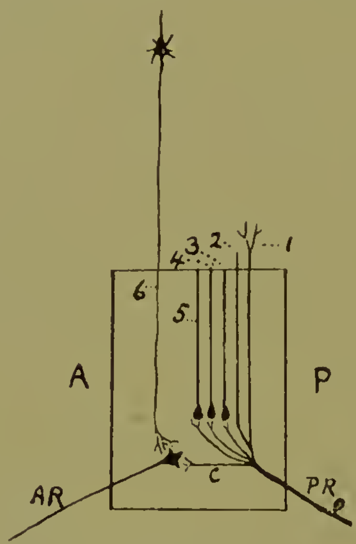


FIG. 43.—Longitudinal Section of Spinal Cord in Antero-posterior direction (diagrammatic).

A = Anterior part.
P = Posterior part.
AR = Fibre of anterior nerve root from cell in anterior horn.
PR = Posterior nerve root, of which the following fibres are indicated:—
C = Reflex collateral passing to nerve cell in anterior horn, and completing the reflex arc PR–C–AR.
(1) Fibre conveying afferent impulses from muscles and joints to the medulla.
(2) Fibre conveying tactile sensation.
(3) Path of afferent impulses from the muscle to the cerebellum by means of a secondary neuron.
(4) Path of afferent impulses for pain and temperature (through secondary neuron).
(5) Path of crossed impulses for touch (through secondary neuron).
(6) Fibre conducting motor impulses from the brain cortex to nerve cell in the anterior horn.

upwards on both sides of the spinal cord, i.e. that the tracts are partially crossed, partially uncrossed. Mann's view is that for tactile sensation all centripetal tracts are open, and that so long as any centripetal tracts are open tactile sensation can be conducted through them, though the chief path of conduction is in the posterior columns.

The tracts conducting the sensations of pain and temperature lie close together. At first these sensations are conducted in the posterior horns of grey matter, but probably they soon cross to the opposite side

of the cord and then pass into the antero-lateral columns in front of the lateral pyramidal tract, and are conducted upwards to the brain, at this region—the spino-thalamic tract. The sensations of pain and temperature are believed by some observers to pass upwards in the antero-lateral tract of Gowers, but this view is strongly opposed by others.

The direct cerebellar tract receives fibres from the cells of the posterior vesicular column of Clarke; the fibres of the former tract pass upwards to the cerebellum. The fibres of the antero-lateral ascending tract also pass upwards to the cerebellum.

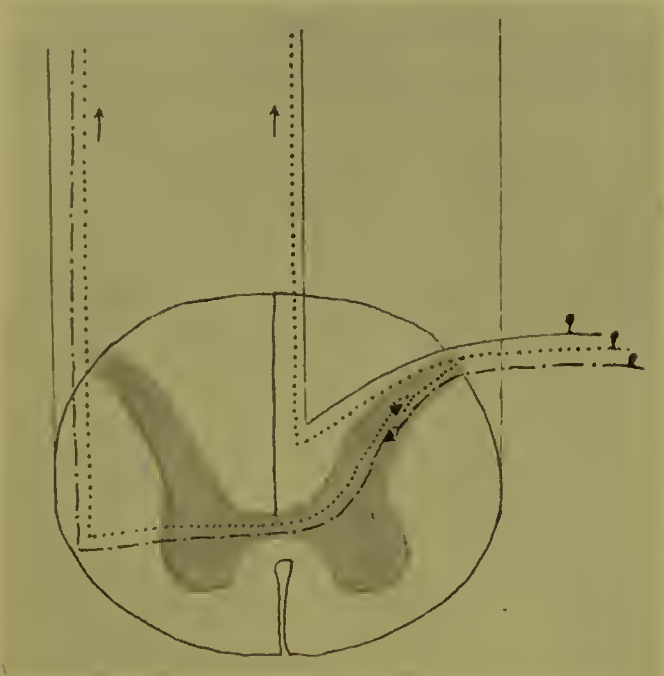


FIG. 44.—Transverse Section of Spinal Cord, showing course of Afferent Impulses.
(1) Those from muscles and joints = continuous line.
(2) Those for tactile impressions = dotted line.
(3) Those for pain and temperature = interrupted line.
Note that impulses (1) do not cross in the Cord. Impulses (2) partly cross and partly pass up on same side. Impulses (3) all cross.

The following views may also be briefly recorded.

Edinger and others think that the tracts conducting sensations of pain and temperature cross in the grey matter of the cord, and that the tracts conveying the sensation of touch and pressure are partly in the posterior columns and partly in the grey matter; the latter cross to the opposite side of the cord whilst the former do not.

Sir W. Gowers thinks that it may be accepted that tracts for the tactile and painful impulses which we feel cross the middle line, soon after entering the cord, but probably some impulses that we do not feel do not cross.

Sherrington states that the sensory tract certainly becomes bilateral a short distance above its entrance into the cord. He adds—"The dissociated forms of anæsthesia occurring in disease, e.g., syringomyelia, and noted experimentally after certain lesions of the cord, indicate that the spinal paths from 'touch spots' and 'cold spots' probably lie together and apart from 'warm spots' and 'pain spots,' which latter probably run together. The latter seem to enter the grey matter very soon after entering the cord, and then to cross in large part to the opposite lateral column. The two former probably do not decussate as soon or to such a high degree."

Some observers believe that the conduction of sensory impulses is chiefly in the grey matter.

According to S. Simpson and P. T. Herring there does not seem to be any specific tract (in the cat) for the conduction of painful sensation excited by immersing the foot in hot water. While any part of the transverse section of the cord remains intact this sensation can pass, though probably the normal path is the lateral column. The only lesion which abolishes all forms of sensation is complete transverse section.

W. P. May and others believe that the fibres subserving touch do not cross so readily as those for pain and temperature.

H. Head and T. Thompson conclude, from a very careful study of the subject, that the spinal cord is the seat of the transmutation of most of the impulses of the peripheral into those of the secondary level of the afferent nervous system. The secondary paths for sensory impulses then cross with greater or less rapidity, so that ultimately all except those subserving the sense of passive position and movemen and tactile discrimination have passed to the opposite side within the limits of the spinal cord. Even these sensory impulses cross after reaching the nuclei of the posterior columns.

The pathological spinal changes in tabes and Friedreich's disease, as well as experiments on animals, appear to indicate that the afferent fibres from the muscles pass upwards in the posterior white columns.

The localization of the pathological changes in anterior poliomyelitis, and the fact that in this disease sensation is unaffected, indicate that the sensory impulses do not pass upwards in the anterior grey matter.

In a case of acute poliomyelitis of the cervical region which the author has recorded, the lesion affected both anterior horns of grey matter and the grey commissures; on one side the posterior horn was also invaded by the inflammatory changes, but on the other side the posterior horn was unaffected. On the side on which the posterior horn was affected there was loss of sensation to pain and temperature whilst tactile sensation was unaffected on either side. But on the side opposite to the analgesia and thermo-anæsthesia, there was invasion of the antero-lateral white matter by the inflammatory changes.

In cases of syringomyelia the sensation for pain and temperature is lost and the lesion affects the posterior horn of grey matter—facts which suggest that the sensations for pain and temperature are conducted at first into the posterior grey matter. In this disease when there is unilateral analgesia and thermo-anæsthesia the lesion is in the posterior horn of the same side (*see* p. 255).

Fig. 44 indicates the course of sensory impulses, which is accepted by most neurologists.

Muscular "Tone."—During life the voluntary muscles are in a state of persistent slight contraction to which the name of "tone" is given. This "tone" is dependent on the integrity of the posterior nerve roots. It is regarded as a reflex tone; it disappears on section of the posterior roots; and is diminished in disease of these roots (as in tabes). The methods of testing muscle tone are described on p. 73.

Reflex Centres.—The spinal cord is not only a conductor of motor and sensory impulses, but in it are situated a series of so-called reflex centres or arcs. Impulses passing to the cord, by afferent nerves, at definite segments excite impulses in the efferent (or motor) nerves of the same segment, and a muscular movement may be produced independent of any voluntary action. The path along which impulses pass between the posterior and anterior nerve roots is known as the reflex

arc, and the portion of it in the grey matter as the reflex centre. "Collateral" fibres from the nerve fibres of the posterior root form one path for the impulses. The end branches of the "collateral" fibres come in contact with the processes of nerve cells of the anterior horn and thus complete the reflex arc (*see* Diagram 45). It is probable that some reflex arcs are completed by intermediate neurons.

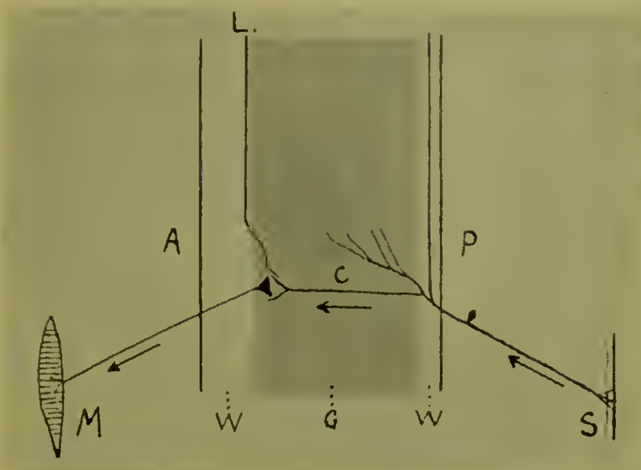


FIG. 45.—Diagram of Longitudinal Section of Spinal Cord, in Antero-posterior direction, showing reflex arc.

A = Anterior.
P = Posterior part.
M = Muscle.
S = Skin.
W = White matter of Cord.
G = Grey matter.
C = Collateral.

L = motor fibre in lateral pyramidal tract from brain to nerve cells in anterior horn. Note path of reflex impulses indicated by arrows from skin S through posterior nerve root, then through collateral (C) to nerve cell of anterior horn, then to muscle M.

The simple reflexes have their reflex arcs in the same spinal segment as that in which the posterior nerve root enters. In complicated reflex movements the arc extends to other segments.

The brain has an inhibitory action on reflex movements, and when this influence is cut off the spinal reflexes are often increased. But the reflexes have been found quite absent below a complete transverse lesion of the cord in man (*see* p. 114). The various definite reflexes which are of importance in clinical medicine are described on p. 62.

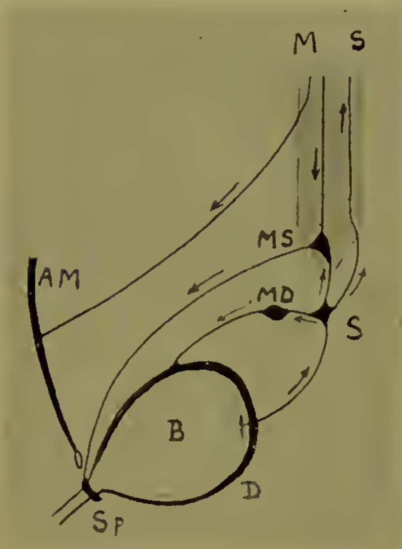
Co-ordination of movement is a function of the brain. The fibres of the posterior median column convey impulses from the muscles and these pass to the cerebellum and thence to the cerebrum. But it is probable that movements in which there is little variation are, to some extent, co-ordinated in the spinal cord.

Visceral Control.—The bladder and rectum are controlled by impulses from the spinal cord which pass through efferent fibres in the anterior nerve roots. At the orifice of both the bladder and the rectum is a sphincter to control the evacuations, and in the walls of each organ are muscular fibres which by their contraction expel the contents thereof.

But the action of both organs is under the control of the will. In diseases of the cord cutting off the cerebral influence, or damaging the reflex centres, this voluntary control is lost, and the bladder and rectum then act involuntarily or in a reflex manner (*see* p. 114) If the sensation of these organs is also lost, the patient is unconscious of the involuntary action.

The reflex centres of the bladder probably consist of a motor part controlling the sphincter; of another motor part controlling the muscular wall of the bladder (detrusor); and of a sensory centre connected with the mucous membrane of the bladder and with the two motor centres. The motor centre for the sphincter and the sensory centre are connected with the brain.

When the bladder is over distended with urine, sensory impulses pass from its walls to the sensory centre, and from this to the motor



AM =Muscles of abdominal wall.
B =Bladder.
D =Muscular wall of bladder (detrusor).
Sp =Sphincter of bladder.
M =Motor impulse from brain to the abdominal muscles and the sphincter.
S =Sensory centre and sensory impulses to the brain.
MS =Motor centre for the sphincter at lower end of spinal cord.
MD =Motor centre for detrusor.

The centres MD and S are in the pelvic sympathetic ganglia according to Müller.

FIG. 46.—Centres for Bladder and Reflex Arcs.

centres and to the brain. But no reflex action occurs in the normal condition unless permitted by the will.

When urine is passed in the normal condition the action of the sphincter centre is voluntarily inhibited, the abdominal muscles are contracted voluntarily, and the tension of urine in the bladder is thus increased; the detrusor centre acts, the walls of the bladder contract, and micturition occurs. The muscular wall of the bladder is said to contract reflexly and not to be under the control of the will.

In the normal condition, when urine is not being evacuated, the sphincter centre is in action.

When the reflex centres are destroyed the sphincter is permanently relaxed, the muscle wall of the bladder is paralysed, and urine constantly dribbles away (*see* p. 115).

Sir W. Gowers has shown, that for the rectum there is a similar mechanism. Two conditions of the sphincter ani may be recognized.

In the normal condition, when the finger is introduced into the rectum there is a relaxation of the sphincter, followed by gentle firm tonic contraction. When the sphincter is paralysed through lesion of the lumbar centre, if the finger be introduced into the rectum there is a momentary contraction (due to local stimulation) followed by permanent relaxation of the sphincter ani.

The functions of the sexual organs in man, and certain functions of these organs in woman (those associated with sexual intercourse), are under the influence of reflex centres, and are also controlled by the will.

The following is regarded as the localization of genito-urinary centres :—

Centre for erection of the penis ,, ,, ejaculation	3rd sacral segment.
,, ,, sphincter ani ,, ,, sphincter of urethra	3rd and 4th sacral segments.

Until recently it was generally accepted that all the reflex centres for the bladder, rectum, and sexual organs were situated at the lower end of the spinal cord.

But according to L. R. Müller, the view that centres for the discharge of urine and fæces are in the lower end of the spinal cord is not correct. From his experiments and observations he concludes that only the ganglion cells for the sphincters of the bladder and anus are situated at this part. The centres for the reflex movements (contraction of the walls of bladder and rectum) as well as for the erection of the penis are believed to be in the pelvic sympathetic ganglia. But this view is not yet generally accepted.

The uterine functions are probably not under the control of the spinal cord, since pregnancy and labour may occur in the normal manner in cases of complete paraplegia.

Trophic Influence.—The motor nerve cells of the anterior horns of grey matter are the trophic centres of the muscles, which they supply with motor nerve fibres. Disease of these cells is followed by paralysis and atrophy of the muscles supplied thereby, with changes in their electrical reactions (*see* p. 74).

The nutrition of bones and joints is probably affected by lesion of the posterior nerve roots, as is indicated by the pathology of tabes. It is believed by many that the posterior roots have also a trophic influence on the skin ; but irritation of the root has more effect than simple loss of function.

Herpes Zoster is caused by an inflammatory lesion of the spinal ganglia of the posterior roots (*see* p. 99). Some observers believe that the pathology of syringo-myelia indicates that the posterior horn of grey matter has a trophic influence on the skin, bones, and joints.

In its broad sense every nerve cell has a trophic influence on some

tissue; and all nerve cells have also a trophic influence on other nerve cells with which they may be connected or related.

Vaso-motor Functions.—According to Edinger tracts for the innervation of blood vessels do not decussate; they probably course in the antero-lateral columns.

Vaso-motor centres are situated in the spinal cord, probably in the intermediate grey matter or in the lateral horn. Vaso-constrictor nerves probably leave the cord in the anterior nerve roots of the dorsal and upper lumbar segments and pass by the rami communicantes to the sympathetic nerves and through these to the vessels. The vaso-dilator fibres also leave the cord in the upper dorsal, lumbar, and sacral regions. Fibres passing to the sweat glands have probably a similar course to the vaso-motor fibres (Obersteiner).

The cervical part of the cord contains fibres which pass to the sympathetic and influence the sugar metabolism in the liver.

In the lower cervical and upper dorsal regions of the cord is the cilio-spinal centre, from which fibres pass to the sympathetic nerves and supply the vessels of the head and face. Fibres from this centre pass through the 1st dorsal (or 1st dorsal and 8th cervical) nerve roots, by the rami communicantes, to the cervical sympathetic and then to the dilator fibres of the pupils. Irritation of the centre, or of fibres coming from it, causes dilatation of the pupil and increase of the palpebral fissure: destruction of the centre or its fibres causes contraction of the pupil and diminution of the palpebral fissure. Fibres also pass from the cervical cord by means of the sympathetic nerves to the heart (accelerator nerves).

Functions of the Lateral Grey Matter.

To the column of nerve cells at the lateral margin of the grey matter, mid-way between the anterior and posterior horns, the term intermedio-lateral tract is given. According to A. Bruce, who has studied its histology thoroughly, it is composed of cells in and near the tip of the lateral horn and of cells at the angle of the formatio reticularis. The tract extends from the eighth cervical to the second lumbar segment. The total number of cells in the tract is very great. Bruce has counted over 88,000 on each side.

All the roots of the spinal cord are connected with ganglia of the sympathetic system by fine non-medullated fibres (grey rami communicantes), but only a limited number by medullated fibres (white rami). The longitudinal distribution of the cells of the intermedio-lateral tract from the 8th cervical to the 2nd lumbar segment corresponds exactly with that of the origin of the white rami of the sympathetic (A. Bruce). According to Gaskell the white rami are efferent.

Section of the cervical sympathetic is followed by degeneration of cells in the intermedio-lateral tract (réaction à distance). Hence A. Bruce thinks it is probable that fibres of the cervical and splanchnic

sympathetic nerves arise from the cells of the intermedio-lateral tracts. The effects of stimulating the various roots of the sympathetic system or the distributions of these fibres are shown in the following table (from the article by A. Bruce); and therefore, these are some (if not all) of the functions of the cells of the intermedio-lateral tract.

Dilatation of Pupil (1) .	1	2	3														
Retraction of Nictitating Membrane: Separation of Eyelids; Protrusion of Eye (1) .	1	**2**	**3**	4	(5)												
Blood-Vessels of Head (1)	(1)	2	**3**	**4**	5												
Secretion of Submaxillary Gland in Cat (1) .	(1)	**2**	3	4	5												
Acceleration of Heart (1)	1	**2**	**3**	**4**	5	(6)											
Hairs of Face and Neck (1)			(3)	4	**5**	**6**	7										
Vaso-Motor Fibres to, and Sweat Glands of Fore-Foot (1) . . .				4	5	6	**7**	8	9	(10)							
Spleen (2).			(3)	(4)	5	**6**	**7**	**8**	9	10	(11)	(12)	(13)	(1)			
Vaso-Constrictors of Portal System (3) .			(3)	4	**5**	**6**	**7**	**8**	**9**	10	(11)						
Viscero-Motor and Viscero-Inhibitory to Small Intestine (4) .						6	7	**8**	**9**	**10**	**11**	**12**	**13**	**1**	2	(3)	(4)
Inhibitory to Small Intestine (5)						6	7	8	**9**	**10**	**11**	**12**	13	1			
Vaso-Motor to Liver, Pancreas, Small Intestine; 1st part of Large Intestine (6) . . .					(5)	6	7	8	9	10	11	**12**	**13**	1	2		
Vaso-Motor to Kidney (7)				4	5	6	7	8	9	10	**11**	**12**	**13**	(1)	(2)	(3)	(4)
Vaso-Motor and Sweat Secretion of Hind-Foot (1)											11	12	13	**1**	2	3	

The numbers within brackets indicate levels where the resulting stimulation was faint, inconstant or doubtful.
The numbers in heavy type indicate levels at which stimulation gave the maximum result.

A Bruce has recorded a case of acute atrophic paralysis with loss of sweating (anidrosis) in the legs. He attributes this symptom and vascular paresis to hæmorrhage and congestion, with subsequent degeneration of nerve cells, which was found in the intermedio-lateral tract (10th dorsal to 2nd lumbar segments). There was in addition degeneration of nerve cells in the anterior horns.

Higier reports three cases of paraplegia (due to meningo-myelitis, hæmorrhage, and sarcoma respectively) in which there was anidrosis in the legs, and a case of cervical syringomyelia with anidrosis in the arms.

Lannois and Porat have published a case of erythromelalgia with gangrene of the limbs, associated with extensive involvement of the intermedio-lateral tract.

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SECTION IV

SYMPTOMS OF DISEASES OF THE SPINAL CORD

Motor Symptoms.

Paralysis.—A common symptom in diseases of the spinal cord is paralysis—the condition in which the patient cannot contract the affected muscles by the influence of the will. A severe lesion affecting both halves of the cord (a transverse lesion) causes paralysis of both legs. To this condition the term paraplegia is given. If the lesion be in the cervical region of the cord, both arms may be affected as well as the legs. When the loss of power in the legs is only partial, the condition is described as para-paresis. When the disease affects one side of the cord only, the paralysis may be unilateral, affecting one limb only (monoplegia), or one group of muscles only (as in infantile paralysis).

According to the condition of the nutrition of the muscles, paralysis may be divided into two forms: (1) Atrophic, (2) Non-atrophic.

In many chronic diseases (as in cancer and phthisis) there is general wasting of muscles; also paralysed muscles may slowly waste, to a *slight extent*, through disuse (simple atrophy—inactivity atrophy). But the term atrophic paralysis is not applied to these cases.

In true atrophic paralysis there is *well marked* and *localised* wasting of the paralysed muscles, which also undergo degeneration. This form of atrophy is distinguished from the simple general atrophy just mentioned by the greater degree of wasting, by its more rapid development, in some cases by the presence of electrical changes (reaction of degeneration), by the loss of the reflexes related to the affected region of the cord, and by the occurrence of fibrillary twitchings, in some cases.

True atrophic paralysis is caused by a lesion of the trophic centre of the muscles (in the anterior horns), or of the nerve fibres between this centre and the muscles affected (i.e. by a lesion of the lower motor neuron). In rare cases it is due to a primary muscular disease (idiopathic muscular atrophy). When the lesion is above the trophic centre usually atrophic paralysis does not occur, though the paralysed muscle may undergo slight simple atrophy from disuse. (To this statement there are a few apparent exceptions.)

There is a very chronic form of degenerative atrophy in which decided paralysis does not occur for a long period. The muscle fibres waste

very gradually; the loss of power simply corresponds to the loss of muscle substance; and hence it is only marked at an advanced period of the disease (as in progressive muscular atrophy).

Motor paralysis may also be divided into two forms according to the condition of the tone of the muscles: (1) Flaccid paralysis, (2) Spastic paralysis.

In flaccid paralysis there is no increased resistance to passive movements in the paralysed parts. In spastic paralysis there is increased resistance (muscular rigidity) to passive movements; the paralysed muscles do not atrophy (except to a slight degree from disuse—simple atrophy): and the reflexes in the affected parts are usually much increased (*see* p. 261).

Spastic paralysis is caused by a lesion of the upper motor neuron (*see* Fig. 17), atrophic paralysis usually by a lesion of the lower motor neuron.

Clonic Spasm.—Spasm, paroxysmal in nature and of short duration, occasionally occurs in spinal disease. Sudden spasmodic contractions of muscles, causing flexion of the legs, often occur in myelitis and other spinal lesions in which there is spastic paraplegia of long duration.

Muscular Contracture.—Distortion or abnormal position of a limb, or segment thereof, caused by persistent tonic muscular spasm is known as contracture (as in the flexed legs in myelitis). The movement at a joint may be restricted or practically abolished through muscular rigidity (muscular spasm).

In spastic paralysis the rigid muscles, and in atrophic paralysis the non-paralysed muscles, may undergo shortening in course of time. When one group of muscles is paralysed, and the paralysis is atrophic, their non-paralysed opponents often undergo shortening. Permanent resistance to passive movement is thus produced.

Abnormal position or restricted movement of a limb, through active muscular spasm or rigidity, should be distinguished from abnormal position through shortening of non-paralysed muscles owing to tissue changes. The former can be removed by passive extension for a few minutes, the latter cannot. The abnormal position from actual shortening of non-paralysed muscles, owing to tissue changes, can be relieved by tenotomy; but in active contraction from muscular spasm tenotomy is useless, as the abnormal condition would soon return.

Primary contracture, through an increase of muscle tone, is known as active or spastic contracture. Secondary contracture, caused by contraction of the antagonists of the paralysed muscles, is known as passive or paralytic contracture.

The Gait of patients suffering from spinal disease is of diagnostic importance and often suggests the nature of the disease. The chief forms of abnormal gait are described under the various diseases—the *spastic* gait on p. 266, the *ataxic* gait on p. 298, the *high stepping* gait on p. 198.

The handwriting is often affected in diseases of the nervous system

owing to various forms of tremor, and to ataxia of the arms as in some cases of tabes, etc.: but most important is the marked irregularity in the handwriting in many cases of early disseminated sclerosis, at a time when the tremor of the hand is very slight, and difficult to detect (*see* p. 281).

Tremor.—Various forms of tremor are observed in nervous diseases.

1. Tremor occurring during repose of the limb, but ceasing or diminishing on voluntary movement with attention—tremor which ceases when an object is grasped or when the hands are held out. The tremor in paralysis agitans is usually of this form.

2. Tremor occurring only on voluntary movement and ceasing during repose (intention tremor). This is the form of tremor in disseminated sclerosis, even at an advanced stage; but several other forms of tremor at a *very early stage*, only occur on voluntary movement.

3. Tremor which occurs during repose, but which is much greater during voluntary movement; as in most cases of marked alcoholic, senile, asthenic, simple and hysterical tremor, and in several other varieties.

In the account of the symptoms in several spinal diseases tremor will be described.[1]

Mechanical Excitability of Nerves and Muscles.—The mechanical excitability of a motor nerve can be tested by tapping it with a percussion hammer, at a point where it is superficial. The result is contraction of muscles supplied by the nerve. In tetany the excitability is so much increased that a light stroking, with the handle of the percussion hammer or with the finger, over the facial nerve causes tetanus of the facial muscles.

The mechanical excitability of muscles is tested by tapping them with a percussion hammer. In the normal condition in many muscles a sharp contraction may be obtained. In cases of paralysis, when the reaction of degeneration is present, the muscular contraction on mechanical stimulation is *sluggish*, like the reaction to galvanism.

The Reflexes.

Changes in the reflexes are of great value in the diagnosis of diseases of the spinal cord.

The more important reflexes are of two kinds:—

1. Tendon and periosteal reflexes, or *deep* reflexes.

2. Skin reflexes, or *superficial* reflexes.

In addition there are other special reflexes—pupillary reflexes and reflexes caused by stimulation of mucous membranes.

Closely associated with the condition of the deep reflexes, both in normal and pathological conditions, is the muscular tone.

In pathological conditions the deep and superficial reflexes may be

[1] For further account of tremor, and the changes produced thereby in the handwriting, see author's article and small monograph—*On Paralysis Agitans and other Forms of Tremor*, Manchester, 1901, and *Medical Chronicle*, 1900–1901.

lost or increased. But often the changes in these two classes of reflexes do not run parallel. Thus, the deep reflexes may be lost, whilst the superficial (or at least certain of them) are present or increased. In tabes dorsalis loss of the knee jerks is often associated with presence of the superficial reflexes. In diabetes mellitus, of the severe form, the knee jerks are often lost whilst the superficial reflexes are present or increased.[1] In Friedreich's disease and other forms of postero-lateral sclerosis loss of the knee jerks is often associated with increase of the plantar reflex, and the latter is usually of the extensor type.

In hemiplegia and in hysterical paralysis the skin reflexes may be lost on the paralysed side, whilst the deep are present or increased.

Whatever may be the exact nature of the deep and superficial reflexes, i.e. whether true reflexes or not (*see* p. 64), the reflex arc must be intact at various definite segments of the cord in order that the reflex may be obtained. The afferent nerve fibres from the part stimulated to the spinal cord, the efferent fibres from the cord to the muscles which contract during the reflex movement, and the connexion between these fibres within the cord, i.e. the completion of the reflex arc, must be uninterrupted when the reflex occurs (*see* Fig. 47). (The connexion between the fibres of the reflex arc entering the cord in the posterior nerve root and the motor cells of the anterior horns of grey matter is probably by the "reflex collaterals.") If the reflex arc is broken down (interrupted) at any part the reflex is lost. But in addition the reflexes may be abolished by inhibitory influences transmitted from other parts of the nervous system, and they may be lost in coma and narcosis. Moreover, the reflexes may be increased by lesions affecting parts of the nervous system outside the reflex arc.

Deep Reflexes.

The most important deep reflexes are the knee-jerk or patellar tendon reflex, the tendo-Achillis reflex, ankle-clonus, the wrist-jerk, and the triceps-jerk.

The Knee-jerk, or Patellar Tendon Reflex.—By striking the ligamentum patellæ (when the leg is in a suitable position) contraction of the quadriceps femoris is produced, sudden extension occurs at the knee, and the lower part of the leg is jerked forward. To this reflex movement the term knee-jerk is applied. In order that the reflex should be produced the leg must be in such a position as to cause slight stretching of the quadriceps muscle, i.e. this muscle must be in a state of passive tension. The knee should be flexed so that the lower part of the leg and the thigh form an angle a little more than a right angle. The reflex is commonly examined when the leg which is being tested is crossed over the other at the knee, the patient being seated on a chair.

When the knee-jerk is not obtained in the ordinary way Jendrássik's

[1] See writer's paper, *Journal of Neurology and Psychiatry*, Oct., 1903.

method of reinforcement should be employed before the reflex is declared to be absent. The patient is seated on a chair, with his feet well in contact with the floor, and the knees bent at an angle a little more than a right angle. He is made to look upwards, to hook his fingers together, and to pull tightly. The observer places one hand on the rectus femoris muscle, and strikes the patellar ligament with a percussion hammer or stethoscope, whilst the patient is counting from 1 to 20. If the slightest knee-jerk be present, a contraction of the rectus muscle will be felt.

When the patient is unable to get out of bed the leg may be raised slightly by the hand of the examiner placed behind the lower end of the thigh, so that the knee is bent at an angle of about 135° ; and with a stethoscope or percussion hammer the ligamentum patellæ is struck.

In whatever way tested, the quadriceps muscles must be relaxed in examining the reflex.

It is probable that the knee-jerk is not a true tendon reflex (i.e. it is not a reflex produced by stimulation of the sensory nerves of the ligamentum patellæ), but is due to muscle reflex action. The knee-jerks can only be produced when the muscle is in a state of slight passive tension and when the blow on the patellar ligament increases the tension. Also it has been shown by experiments on animals, that after division of all the sensory nerves connected with the patellar tendon the knee-jerk can still be obtained. Further the time between the striking of the ligament and the jerk of the limb is too short for the occurrence of a true nervous reflex.

It is probable that the knee-jerk is dependent on muscle reflex irritability or muscle " tone " : and this is generally believed to be dependent on the reflex arc being intact. Hence if the knee-jerk be regarded as a muscle reflex, the reflex arc of nerve fibres or nerve tracts, connecting the muscle with the cord, must be intact in order that the jerk shall occur, i.e. the arc shown in Fig. 47 must be uninterrupted in order that the knee-jerk shall be obtained. As Sherrington states: " the knee-jerk though not itself a reflex, is a delicate index of the reflex spinal tonus. It is a simple spasm of part of the quadriceps extensor muscle, usually elicited by a tap or other brief mechanical stimulus applied to the muscle fibres mediately through the tendon." The reflex is obtainable only from muscle fibres possessed of their " neural " tonus.

The reflex arc which must be intact, in order that the knee-jerks may occur, is situated at the level of the second, third and fourth lumbar nerve roots.

Loss of the knee-jerk, or Westphal's sign, occurs in many diseases, and is a sign of great diagnostic value.

In health the knee-jerks are always present, but occasionally it is difficult to elicit them, and Jendrássik's method may be necessary.

The knee-jerks may be *lost* in the following conditions :—

1. In lesions of the reflex arc in its afferent part, as in tabes dorsalis

and Friedreich's disease, in peripheral neuritis from alcohol, diphtheria, arsenic, diabetes and other affections. Lesion of the posterior root fibres directly after they enter the cord is probably the cause of the loss of knee-jerks in some cases of diabetes mellitus and brain tumour (*see* p. 37). In spinal meningitis and tumour of the meninges affecting the lumbar region the knee-jerks may be lost from lesion of the posterior nerve root.

2. In lesions of the grey substance of the anterior horns at the level of the reflex arc (Lumbar 2, 3, and 4 segments) as in poliomyelitis, acute, subacute and chronic.

FIG. 47. — Reflex Arc, which must be intact in order that the knee-jerk shall occur. At lower part of figure femur, tibia, and patella, are indicated in deep black. Rectus muscle shaded with parallel lines. Dotted line = afferent fibres from muscle, entering the cord by the posterior nerve root. These impulses are transmitted to the anterior horn by collaterals (C). From the nerve cells of the anterior horn the impulses pass to the muscle by the motor nerve (shown by a continuous line).

3. In lesion of the anterior nerve roots or peripheral fibres of the motor nerves, as in spinal meningitis or tumour of the meninges, and in peripheral neuritis of various forms.

4. In lesions destroying the cord at the upper lumbar region—myelitis, compression myelitis from various causes, softening, hæmorrhage, tumour, and occasionally disseminated sclerosis.

5. In *complete* transverse lesion of the cord above the lumbar region—myelitis, hæmorrhage, tumour, compression myelitis, etc. — the knee-jerks may be lost though the reflex arc is intact (*see* p. 115). The knee-jerks may be lost *for a few days* after an *incomplete* transverse spinal lesion when the onset of the lesion is abrupt.

6. In advanced pseudo-hypertrophic paralysis and idiopathic muscular atrophy the knee-jerks may be lost.

7. In coma and in the narcosis produced by anæsthetics.

8. In some cases of tumour of the cerebellum, of the frontal region and of other parts of the brain, and sometimes in cerebral meningitis, the knee-jerks are lost. (In intra-cranial tumours there is often degeneration of the intramedullary fibres of the posterior roots in the lumbar region, directly they have passed through the pia mater.)

9. The knee-jerks are lost in some cases of general paralysis of the insane, and they are often absent, on one or both sides, in cerebral hæmorrhage, or softening, for a short time at the onset of the affection.

10. "Extreme bodily fatigue diminishes, and occasionally abolishes

5

the knee-jerk for a while" (Sherrington). B. Abrahams has pointed out that in cases of sore throat the loss of the knee-jerks is a point in favour of true diphtheria. The knee-jerks are sometimes lost in acute pneumonia.

The knee-jerks are *increased* when there is secondary or primary degeneration of the lateral pyramidal tracts.

(*a*) They are increased in cases of sclerosis in these tracts below a transverse lesion of the cord (usually incomplete) in the dorsal or cervical region; as in descending sclerosis secondary to transverse myelitis, disseminated myelitis, compression myelitis (from caries, spinal tumour, and other causes), spinal hæmorrhage, syringomyelia, spinal or meningeal tumour, meningitis, and some forms of spinal syphilis.

(*b*) The knee-jerks are increased in many diseases or degenerations affecting the lateral pyramidal tracts; as in amyotrophic lateral sclerosis, ataxic paraplegia, combined postero-lateral sclerosis, primary lateral sclerosis, hereditary lateral sclerosis, Erb's spinal syphilitic paralysis, and disseminated sclerosis.

(*c*) The knee-jerk is increased, in course of time, on one side—the side opposite to the lesion—in cerebral hæmorrhage, thrombosis, embolism, tumour, or abscess.

(*d*) Sometimes the knee-jerks are much increased in general paralysis of the insane.

(*e*) The knee-jerks are often increased in neurosis, neurasthenia, and hysteria, after large doses of strychnine, in uræmia, and in tetanus.

When the reflex excitability is increased a tibial periosteal reflex may often be obtained. Tapping the inner surface of the tibia produces a contraction of the thigh muscles, especially the quadriceps. In similar conditions of increased excitability a crossed reflex may be sometimes obtained—when the ligamentum patellæ of one side is tapped a contraction of the quadriceps muscle occurs, not only on the same side, but also on the opposite side.

When the reflex excitability is much increased patellar clonus may be obtained, i.e. when the patella is suddenly jerked downwards, by the index finger and thumb placed on its upper border a series of clonic contractions of the quadriceps muscle occurs, and oscillating movements of the patella are produced.

Simple increase of the knee-jerks, without other symptoms, is of little importance; but if ankle clonus, patellar clonus, Babinski's reflex, or rigidity of the legs be also present, then the increase of the knee-jerks is of diagnostic importance.

The tendo Achillis jerk. This reflex is obtained by striking the tendo Achillis sharply close to its insertion into the os calcis. In the normal condition the blow is followed by a sharp contraction of the calf muscles and a plantar flexion movement of the foot at the ankle joint. The reflex is easily obtained when the patient kneels on a chair with the foot projecting a few inches over the edge of the chair seat. The muscles of

the leg should be completely relaxed when the reflex is being tested. The following method is useful : The patient stands on one leg by the side of the chair, he faces the chair back, and places the other leg on the seat of the chair in the kneeling position with the foot projecting a little over the edge. It is also advisable for the examiner to place his left hand on the lower part of the leg and to press the leg downwards on to the chair, so that the calf muscles may be relaxed. With a stethoscope or, better, a *heavy* percussion hammer, held in the examiner's right hand, the tendo Achillis is struck just above the heel. It is better to strike the tendon a little to its outer side. When the patient is in bed the Achillis-jerk may be obtained in the following manner : the patient turns on to his side; the knees are slightly flexed; the anterior part of the foot is pressed upwards by the examiner and the tendo Achillis struck with the percussion hammer.

The tendo Achillis jerk is constantly present in healthy persons under fifty years of age. E. Bramwell states that it is sometimes absent in healthy persons over the age of fifty. But other observers have found the reflex always present in health, and this has been my own experience. (Thus von Sarbó found it always present in 300 persons not suffering from any nervous disease.)

In those affections in which the knee-jerks are lost the tendo Achillis jerks are usually also absent, as in tabes, peripheral neuritis, in some cases of diabetes mellitus, in certain cases of general paralysis of the insane. But the tendo Achillis jerk sometimes disappears before the knee-jerk. In several cases of very early tabes I have found the tendo Achillis jerk lost for some months before the knee-jerks have disappeared. At the onset of peripheral neuritis I have found the tendo Achillis absent for a very short time when the knee-jerks have been present. At a later period the knee-jerks have also disappeared.

In some cases of diabetes mellitus I have found the tendo Achillis jerks absent when the knee-jerks have been present (*see* p. 371). The tendo Achillis jerks were lost in 19 out of 50 cases of diabetes ; in 8 of these 19 cases the knee-jerks were also lost, in 11 they were present.

In sciatica the tendo Achillis jerk is often lost on the affected side, but is present on the healthy side.

Sometimes the tendo Achillis jerk is lost after syphilis, when no other signs of tabes or general paralysis are present.

The tendo Achillis jerk will be absent like the knee-jerk in lesions of the lumbar and sacral regions (transverse myelitis, tumour, poliomyelitis, etc.), which break down the reflex arc on the integrity of which these jerks depend. In lesions of the cauda equina the tendo Achillis reflexes are lost, whilst the knee-jerks are preserved. In infantile paralysis affecting the calf muscles on one side, I have found the tendo Achillis jerk lost on the diseased side, but present on the opposite side : the knee-jerks were both present.

In alcoholic heart-failure I have found the tendo Achillis jerk usually

lost (on both sides in 76 per cent. of the cases, on one or both sides in 85 per cent.) even when other signs of peripheral neuritis have been absent or very slight. Often, but not always, the knee-jerks have been present.

In cases of cardiac failure,[1] or dilatation, I believe that the loss of the tendo Achillis reflexes is a sign in favour of the heart affection being due to the taking of excessive quantities of alcoholic beverages, other causes of the loss of the tendo Achillis jerk being excluded. The reflex, however, is not always lost in slight cases.

Rolleston has shown that the tendo Achillis jerk is often lost in diphtheria, even at an early period of the disease. At the early stage of general paralysis of the insane the tendo Achillis reflexes may be lost whilst the knee-jerks are present.

Ankle-clonus.—When the reflex excitability is much increased, sudden jerking of the foot upwards, by the hand placed under the anterior part of the sole, is followed by clonic contractions of the calf muscles and a series of oscillations of the foot. This is known as ankle-clonus. The knee should be supported by the hand posteriorly, and should be slightly flexed (passively), when examination for ankle-clonus is made.

Ankle-clonus is a sign of increased reflex excitability and is almost always an indication of organic disease of the brain or spinal cord—of a lesion of the upper motor neuron.

A slight ankle-clonus is sometimes seen in hysteria, but the jerks are few and the clonus is not sustained. I have seen slight ankle-clonus temporarily, in cases of acute rheumatism, and other joint diseases, in which the ankle joints had been affected.

Reflexes of the Arm.—Many reflexes can be obtained in the arms by striking the tendons of various muscles, providing the muscles be in a state of passive tension. Some of these reflexes are not always obtained in health, and this fact impairs their diagnostic value. Increase or loss of these reflexes on one side is, however, of some diagnostic importance.

The chief arm reflexes are : the triceps-jerk, the wrist-jerk, and the scapulo-humeral reflex.

The triceps-jerk can be obtained by allowing the arm to hang loosely over the back of a chair, the upper arm being supported by the chair back just above the elbow. The elbow should be bent at an angle a little less than a right angle. In this position the triceps is placed in a condition of passive tension. The triceps tendon is struck with a percussion hammer just above its insertion into the olecranon. In the normal condition the triceps contracts and the elbow is extended (the forearm being jerked away from the chair back).

Some observers state that the triceps reflex is always present in health.

[1] In 21 cases of alcoholic heart-failure the tendo Achillis jerk was lost on both sides in 18 ; on one side in 2 ; it was present on both sides in 3 (but these were slight cases).

In 100 cases of heart disease, not due to alcoholism, this reflex was present in 98 and lost in 2 only, and in both of these early tabes was very probable.

Attention should be directed to the triceps muscle, as well as to the movements of the forearm ; and since a feeble contraction of the long head of the triceps may occur without producing any movement of the arm, it is well to feel the muscle with the finger so that the slightest contraction may be detected.

The wrist-jerk, or radial periosteal reflex, is obtained by striking the lower end of the radius, at the region of the styloid process, with a percussion hammer, the elbow being flexed at an angle a little more than a right angle and the forearm being midway between pronation and supination. The reflex movement obtained consists in flexion of the elbow with slight pronation of the forearm. The wrist-jerk cannot always be obtained in health. In 100 healthy men examined by the writer the wrist-jerk was absent in 26. The movement is chiefly produced by contraction of the biceps and supinator longus.

The wrist-jerk is lost in progressive muscular atrophy (Duchenne-Aran type), but is increased in amyotrophic lateral sclerosis : it is lost in Erb's paralysis of the arm on the affected side (paralysis of deltoid, biceps, and supinator longus from lesion of the fifth, or fifth and sixth, cervical nerve roots) : it is lost in the upper arm type of progressive muscular atrophy when the forearm and hand muscles have not become affected : it is often present in lead paralysis, but usually lost in alcoholic paralysis when the arms are affected.

When the forearm is pronated and supported, and the hand allowed to hang down at the wrist, a reflex extension of the hand may be obtained, in certain diseases, by striking the extensor carpi radialis or the extensor carpi ulnaris just at their insertion into the bases of the metacarpal bones. These reflexes are frequently absent in health, but are often well marked when there is spastic paresis of the arm. In disseminated sclerosis and other affections the presence of these reflex on one side, whilst they are absent on the other side, is a point of some value.

A scapulo-humeral reflex has been described by Bechterew. It is obtained by striking the inner margin of the scapula with a percussion hammer near its lower angle (not at lower angle).

The reflex movement is due to adduction of the humerus, often there is also slight external rotation ; and sometimes the movement consists of abduction of the arm and flexion of the elbow. In testing this reflex, Steinhauser recommends that the trunk should be bent forwards and that the arms should hang down. By many authors the reflex is said to be constantly present in health.

Superficial or Skin Reflexes.

The more important of the superficial reflexes are the plantar, cremasteric (or inguinal), abdominal and epigastric reflexes.

If the skin reflexes are present the spinal reflex arc must be intact : but absence of a superficial reflex is not conclusive of a lesion in the

reflex arc, since cerebral lesions may cause these reflexes to disappear, even when the spinal arc is normal.

The superficial reflexes are sometimes, but not always, increased when a lesion above the reflex arc cuts off the inhibitory cerebral influence. They are also increased when there is hyperæsthesia of the skin, and when the excitability of the reflex arc is increased, as in tetanus.

The superficial reflexes are diminished or lost when there is anæsthesia or paralysis caused by lesion at some part of their reflex arc (cord or peripheral nerve), and they may be lost when the excitability of the reflex arc is diminished as in coma. In hemiplegia, from cerebral hæmorrhage or softening—the epigastric, abdominal, and cremasteric reflexes of the paralysed side may be diminished or lost. Directly after an attack of apoplexy the superficial reflexes of both sides may be lost. As already mentioned, the condition of the superficial reflexes does not run parallel with that of the deep reflexes. When the latter are lost the former may be present or increased (as in some cases of tabes and diabetes mellitus).

A complete transverse lesion of the cord may cause loss of both the tendon and superficial reflexes below the lesion.

The **epigastric reflex** is obtained by stroking—from without downwards and inwards—the side of the thorax at its lower part, with the finger, or the handle of a percussion hammer. By stimulating the skin in this manner retraction of the epigastrium is produced. The **abdominal reflex** is obtained by stroking the side of the abdomen at its upper part from without downwards and inwards towards the middle line ; retraction of the middle of the abdomen is produced, and the umbilicus is drawn to the stimulated side. These two reflexes are not always present in health ; they are often absent when the subcutaneous tissue of the abdominal wall contains much fat, or when the abdominal wall is very flaccid. According to Strümpell the abdominal reflexes are so often absent in disseminated sclerosis that their loss is of some diagnostic value. The reflexes may also be lost in Friedreich's disease.

The **cremasteric reflex** consists in a contraction of the cremaster muscle, and retraction of the testicle on one side, when the innner surface of the thigh is stroked with the finger nail or the handle of a percussion hammer. Geigel has pointed out, that in the female stimulation of the inner surface of the thigh, in the same manner, causes a strong contraction of the lowest bundle of the internal oblique muscle just above Poupart's ligament (87 per cent. of cases examined).

The **plantar reflex** is obtained by lightly stroking the sole of the foot ; flexion of the outer toes and finally of the great toe occurs ; also the whole foot may be dorsi-flexed when the reflex movement is strong.

Babinski has shown, that in the normal condition, when the plantar reflex is obtained, the toes are flexed (i.e. bent towards the sole of the foot). But in certain diseases an opposite movement of the toes occurs, (i.e. the toes are extended towards the dorsum of the foot) : and the

extension is especially marked in the great toe. This is known as the extensor type of plantar reflex or Babinski's reflex. According to Babinski the extensor type of plantar reflex indicates lesion of the crossed pyramidal tract in some part of its course; it is the earliest sign of such a lesion; it never occurs in the normal adult, and it does not occur in functional affections of the nervous system. In children under the age of three years an extensor type of plantar reflex is obtained; after that age it is exceptional in children.

In the following table Collier gives the chief characteristics of the three forms of plantar reflex:—

	Normal Flexor Type.	Extensor Type.	Infantile Response.
Movement	Quick	Often deliberate	Quick
Muscle first to contract with a minimal stimulus	Tensor fascia femoris	Extensor prop. hall.	Ext. of toes
Certainty of response to each stimulus	Less certain	Certain	Less certain
Position of toes . . .	*Flexion*: adduction	*Extension*, spreading	Extension, spreading
Obtained more easily by stimulating	Inner part of the sole	Outer part of the sole	—
Movement of ankle . .	Dorsi-flexion and inversion both conspicuous	Dorsi-flexion and inversion both slight	Dorsi-flexion and inversion both conspicuous

The plantar reflex is better obtained when the sole of the foot is dry and warm. In testing for the reflex a minimal stimulus should be applied to the sole by some hard object, such as a penholder or the end of a stethoscope, whilst the patient's attention is distracted. The sole of the foot should be stroked gently from behind forwards. A movement of dorsal flexion of the ankle should not be mistaken for extension of the toes.

The normal flexor type of plantar reflex occurs only when the spinal reflex arc (including the peripheral nerves), is intact, and when also the fibres of the lateral pyramidal tract are *not* affected.

When the fibres of the lateral pyramidal tract are diseased (or when, in the infant, they have not fully developed), the plantar reflex is of the extensor (Babinski) type. When the reflex arc is broken down the plantar reflex is lost. Thus it is lost in lesions of the peripheral nerves, and of the spinal cord at the upper sacral region (*see* Fig. 48).

In almost all cases of lesion of the lateral pyramidal tract the extensor type of plantar reflex is obtained. It is one of the first signs of such a lesion, and is the last to disappear when the lesion is temporary.

In hysteria and functional diseases the plantar reflex is sometimes lost or difficult to obtain: but when elicited it is of the flexor type. The type of plantar reflex is very useful in the diagnosis between hysteria and disseminated sclerosis. In the latter disease the extensor type of plantar reflex is common. Whenever the extensor type of reflex

is obtained the disease is not purely functional, and this form of reflex is sometimes the first clear objective sign of organic disease.

The extensor type of plantar reflex is met with in all affections causing spastic paraplegia or paraparesis (*see* p. 263). It occurs on the paralysed side shortly after an attack of cerebral hæmorrhage: it is present on the paralysed side in hemiplegia. It may also occur in uræmia (S. Barnes) and in many cases of coma from brain disease.

It is present in Friedreich's disease and in various forms of postero-

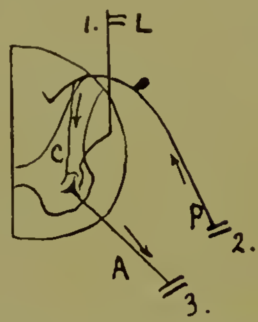


FIG. 48.—Diagram of one-half of the Spinal Cord, indicating the lesions which produce abnormal conditions of the Plantar Reflex.

P = Posterior nerve root and sensory peripheral nerve.
A = Anterior nerve root and motor nerve.
C = Reflex collateral.
L = Motor fibre of lateral pyramidal tract conveying impulses to nerve cell in anterior horn.
P, C, A = Reflex arc for plantar reflex.

A lesion at 1 will cause the reflex to be of the extensor type (Babinski's reflex), whilst a lesion at 2 or 3, or at C, will interrupt the arc and abolish the plantar reflex.

lateral sclerosis, although the knee-jerks may be absent. In the following diseases, if the plantar reflex can be obtained it is of the normal flexor type: tabes, peripheral neuritis, neurasthenia, paralysis agitans, chorea, and poliomyelitis. In cerebral diplegia with choreiform movements the plantar reflex is of the extensor type, whilst in ordinary chorea it is of the normal flexor type.

Oppenheim's Reflex.—In the normal condition, when the handle of a percussion hammer or the ball of the thumb is drawn, in a forcible manner from above downwards along the inner surface of the leg (below the knee), either a plantar flexion of the toes occurs or there is no reflex movement. In cases of spastic paralysis extension of the toes often occurs. I have obtained this reflex in a few cases of early spastic paraplegia (due to vertebral caries, etc.) before the Babinski type of plantar reflex could be obtained.

Bulbo-Cavernous Reflex.—When the dorsum of the glans penis is pinched or pricked, a contraction of the bulbous part of the urethra can be felt by the finger pressed upward on the urethra behind the scrotum. When this reflex is present we may conclude that the reflex arc is intact at the level of the third sacral segment. The reflex is often lost in tabes.

Paradoxical Contraction.—In several nervous diseases a peculiar

symptom is sometimes observed to which this name has been given. When the distance between the origin and insertion of a muscle is suddenly diminished a contraction of the muscle occurs. Thus on passive dorsi-flexion of the foot a tonic contraction is excited in the tibialis anticus muscle. This symptom has been observed in rare cases of disseminated sclerosis, paralysis agitans, and tabes.

The pilo-motor, or **goose-skin reflex** of J. Mackenzie (of Burnley). By stroking the skin, or by the application of the faradic current, a reflex goose skin appearance may be produced—the pilo-motor reflex (*see* J. Mackenzie's papers, *British Med. Journal*, 1906, June 23, and *Brain*, 1893, p. 515).

Tibialis phenomenon of Strümpell.—In lesions of the crossed pyramidal tract, Strümpell has pointed out, that when the lower limb is flexed at the hip and knee an involuntary dorsi-flexion of the foot with inversion occurs. The marked contraction of the tibialis anticus can be seen and felt. This sign is best marked when the limb is flexed against resistance made by the examiner's hand placed on the patient's knee

Muscle Tonus.

The condition of the muscle tone should be ascertained along with the state of the reflexes.

As already mentioned, persistent increased muscular tone, hypertonus (rigidity or tonic spasm), occurs in spastic paralysis—when the crossed pyramidal tracts of the cord are sclerosed or irritated.

The increased tone or rigidity is detected by the tense condition of the muscles and is felt on attempting to perform passive movements of the limbs (*see* p. 265).

In spastic paralysis increase of the tendon and periosteal reflexes is usually associated with increase of the muscle tone.

Rigidity may also be produced by irritation of motor nerve roots in various diseases. In most cases the condition is chronic, but in cases of meningitis, acute tonic spasm, or muscular rigidity frequently occurs.

Fig. 49.—Method of measuring degree of Hypotonus.

The tone of the muscles is diminished in some affections, so that

the resistance to passive movements is much less, and the extent of the movements greater, than in the normal state; also active movements can be performed to a greater extent than in the normal condition. This diminished tone, atony, or hypotonus is met with in cases of flaccid paralysis, and also in tabes dorsalis and some other diseases in which the affected muscles are not paralysed.

In tabes active movements can often be carried out to an abnormal extent. In health, when the trunk is in the horizontal position, the leg can only be raised to an angle of 65–75°, if the knee be kept fully extended. But when hypotonus is present, as in tabes, the leg can often be raised to 90° or more (the knee being kept fully extended).

I have used a small graduated semicircle—with a long piece of string fixed to the middle point of the diameter of the semicircle—for the estimation of the degree of hypotonus. The semicircle is held in contact with the horizontal surface of the couch on which the patient rests, the string is held stretched and parallel with the raised and extended leg. The angle to which the string is raised is read off on the graduated semicircle (*see* Fig. 49).

Hypotonus is often associated with loss of the knee-jerks and deep reflexes, as in tabes.

Spastic and Atrophic Paralysis.

The chief features of two important forms of paralysis, spastic and atrophic, have been often referred to. It may be of service to repeat these features in tabular form.

Lesion of Upper Motor Neuron: *Spastic Paralysis.*	Lesion of Lower Motor Neuron: *Atrophic Paralysis.*
1. Rigidity of muscles	Muscles flaccid.
2. No atrophy of muscles (except a little from disuse)	Well-marked atrophy of paralysed muscles.
3. Deep reflexes present and usually increased: ankle-clonus often present	Deep reflexes often absent; ankle-clonus absent.
4. If leg affected, plantar reflex of extensor type (Babinski reflex)	Plantar reflex absent or of normal type (unless flexors of toes paralysed).
5. No reaction of degeneration on electrical examination	In many cases reaction of degeneration, partial or complete.

Ataxia.

In the normal condition, in the performance of any movement there is a combined action of certain muscles. The contraction of these muscles occurs to a suitable extent, in a suitable direction, and with a suitable force, i.e., a number of muscles are co-ordinated in their action. (1) A number of muscles may act together, or be simply co-ordinated in producing a definite movement. (2) In other movements a number of simple muscular contractions follow one another—complicated co-ordination.

When combined, regulated, or co-ordinated action does not occur, ataxia is the result.

Co-ordination centres are situated in various parts of the central nervous system.

For the suitable distribution of motor impulses, sensory impressions are necessary—sensations of movement, of resistance, of weight, sensations from the joints and skin, etc. Impulses from the eye and ear aid in co-ordination. When the afferent impulses are defective the motor impulses are not properly co-ordinated and ataxia occurs.

It is important to remember that the muscular power may be little affected or unimpaired in the ataxic limb, and that ataxia is quite different from paralysis or paresis.

Disturbances of co-ordination or ataxia occur (*a*) when the co-ordination centres (in the cerebellum or cerebrum) are affected, or (*b*) when there is a lesion of the afferent tracts going to these centres.

In the latter group of cases the lesion may be in the afferent fibres coming from the muscles, and the seat of the lesion is often in the posterior nerve root fibres. In such cases, when the lumbar nerves are affected, the knee-jerks are often lost. In other cases the afferent fibres are affected in the posterior median columns of the cord; in these cases the knee-jerks may be present.

Co-ordination of the action of muscles is necessary, not only in various movements, but also for the maintenance of the equilibrium of the body. The centres for the latter are, probably, in the cerebellum. Afferent impulses are probably conveyed to the cerebellum from the eyes and the semicircular canals, as well as from the muscles, joints, and skin.

There are two important forms of ataxia—that common in tabes, and that characteristic of cerebellar disease.

In the ataxia of *tabes*, in walking the legs are moved irregularly; the extent and the force of the muscular contractions for the object of the movement are excessive; the limbs deviate laterally from their normal course; the heels are placed down on the ground with a stamp, and the legs are well separated, especially in advanced cases. The body is held stiff, the eyes are directed down to the feet; when the eyes are not directed to the feet, or when the eyes are closed, the ataxia is more marked. At the early stage the ataxia is only noted when the eyes are closed. There is ataxia on standing—static ataxia—in marked cases; but at an early stage this is seen only when the eyes are closed (Romberg's sign). There is ataxia or inco-ordination of the movements of the limbs when the patient is sitting or lying down. This is observed when the patient attempts to touch one knee with the heel of the opposite leg, or to touch the nose tip with the index finger, or to bring the tips of the index fingers together. The legs are usually more affected than the arms.

In the inco-ordination of *cerebellar* disease the ataxia consists chiefly

in inability to maintain the equilibrium of the body in standing and walking. The gait is uncertain, reeling and staggering; the body deviates from side to side in a zig-zag manner; and the feet are widely separated. The gait and attitude are like those of a drunken man. There is uncertainty rather than excess of movement. The ataxia is usually not increased when the eyes are closed. When the patient is sitting, or lying in bed, the separate movements of the limbs are usually not ataxic. In walking there is often a tendency for the legs to go forwards and the body to fall backwards (Babinski). Often there is a tendency to fall to one side, or to fall forwards, or backwards.

The ataxia of the combined postero-lateral degenerations and scleroses of the cord (such as occur in association with severe or pernicious anæmia, cachexia, syphilis, various toxic conditions, and in ataxic paraplegia) corresponds to that of tabes. The ataxia of Friedreich's disease and that of certain rare cases of disseminated sclerosis have the characters of cerebellar ataxia; but in addition there is ataxia of separate movements as in tabes (démarche tabéto-cérébelleuse of Charcot). The cerebellar form of ataxia is met with in tumour, abscess, sclerosis (disseminated sclerosis), and general atrophy affecting the cerebellum.

Ataxia may thus be caused by lesion of various parts of the nervous system—cerebellum, cerebrum, medulla, pons, spinal cord and peripheral nerves. (The chief spinal affections causing ataxia are mentioned on p. 291. The testing of ataxia on p. 298.)

Sensory Symptoms.

In spinal diseases sensation is very often less affected than motor power. When a lesion involves both motor and sensory structures, sensation is often affected to a much less extent than motion, and this fact suggests that the sensory fibres are less readily damaged than the motor by diseased processes. (Head's views on the forms of peripheral sensation are given on p. 103.)

Sensory symptoms may be divided into two groups, objective and subjective.

I. Objective Sensory Disturbances.—The general methods of examination of sensation do not fall within the scope of this work, but it may be here mentioned, that in testing sensation, careful attention is necessary on the part of the patient, and much patience on the part of the medical man. Also, several repetitions of the examination are desirable. The objective sensory disturbances of chief importance in spinal disease are: loss of sensation to tactile impressions—tactile anæsthesia; loss of sensation to pain—analgesia; loss of the sensation for temperature—thermo-anæsthesia; affections of the "muscular sense;" and loss of the vibrating sensation.

1. Sensation to *tactile* impressions may be tested by touching the skin lightly with the finger, with a fine brush, with a small piece of

cotton wool, or with the head of a pin, the patient's eyes being closed. On parts of the skin where hairs can be seen the tactile sensation can be well tested by bending the hair sharply, by means of a quick stroke with the point of a pencil, in the opposite direction to that to which it is inclined. It is important that the patient's careful attention should be given to the examination; he should be asked to say "now" when he feels the touch, or to indicate the place touched. The sensation on corresponding parts of the body should be compared on each side.

In spinal diseases tactile sensation is sometimes completely lost—anæsthesia; it may be diminished—hypæsthesia; or increased—hyperæsthesia. Usually hyperæsthesia is associated with increased sensation to pain—hyperalgesia, and ordinary tactile sensation may then produce pain. In mapping out areas of anæsthesia, it is better to test the anæsthetic part first, and to pass from this to the part where sensation is normal.

When the cutaneous sensibility is impaired, but not lost, a pair of compasses, or the æsthesiometer, may be used for measuring the relative sensibility of the skin. In health the points of a pair of compasses, placed in contact with the skin, can only be felt as two impressions when they are a certain distance apart. When nearer together they are felt as one impression. The distance to which the points of the compasses must be separated, in order to produce two impressions, varies in different parts of the body; at the tip of the tongue it is 1 mm.; on the lip, 4 mm.; on the chest, 11 mm.; on the neck, 52 mm.; on the ball of the thumb, 9 mm. (E. H. Weber). The æsthesiometer may be used in making these measurements. Two metal points slide along a graduated metal bar and their distance apart, when they cause two impressions, can be read off from the millimetre scale. In cases in which the cutaneous sensibility at any region is impaired, but not lost, the points of the compasses, or of the æsthesiometer, must be more widely separated than when sensation at that region is normal, in order that two impressions may be felt; and this distance, compared with the normal distance, gives an indication of the degree of diminution of sensation. It is important to remember, that when the two points are separated in a direction corresponding to the long axis of the limb they are less readily distinguished as two points than when separated in the transverse direction. Also by practice the power of distinguishing the two points is increased. For these and other reasons æsthesiometric measurements are not of great clinical value.

Localisation of Tactile Sensation.—The ability to localise tactile impressions is tested by touching the skin in various parts of the body, when the patient's eyes are closed, and asking him to name the point touched, or to put his finger on the spot. The patient will be able to do this in the normal condition; in some diseased conditions he may be able to feel the impression, but may not be able to localise it correctly—topo-anæsthesia.

Two other symptoms require mention here. In the condition known as polyæsthesia, when the skin is touched with one object, two or more impressions are felt. In allocheiria, when one side of the body is touched, an impression is not felt on that side, but on the other side. Both of these symptoms are sometimes present in tabes.

2. The sensation for *pain* may be tested by the prick of a needle or pin, by pinching the skin, or by the faradic current. The sensation to pain may be diminished (hypalgesia) or lost (analgesia), or increased (hyperalgesia). Also, the sensation for pain may be delayed (as in tabes). Thus, when the patient is pricked with a pin, and told to say "now" directly the painful sensation is felt, a short interval of time elapses between the prick and the painful sensation it produces.

3. Sensation for *temperature* may be tested by applying to the skin test tubes containing hot or cold water. (Fragments of ice may be placed in the tube instead of cold water.) A useful method for rapid examination is to breathe upon the patient's skin at a very short distance, as a test for the sensation of warmth, and to blow on to the skin at a greater distance as a test for the sensation of cold. The sensations for both heat and cold are not always affected in the same case, hence both of these forms of sensation should be tested. Strong temperature sensations (especially warmth) produce pain. When there is anæsthesia for cold only, touching the skin with a piece of ice not infrequently produces a sensation of warmth. (Perverse temperature sensation—Strümpell.) The opposite condition, in which warm objects cause a sensation of cold, is much rarer.

Changes in the temperature sense are frequent in diseases of the spinal cord. Loss of sensation to temperature—thermo-anæsthesia—may be present when other forms of sensation are normal (as in some cases of spinal syphilis). Hence, in all cases, the sensation for temperature should be examined before concluding that sensation is unaffected: and it is important to examine the sensation for both cold and heat. I have found loss of sensation for cold when sensation for warmth and all other forms of sensation have been normal (as in some cases of spinal syphilis).

The condition, in which there is loss of sensation to pain and temperature whilst tactile sensation remains normal, is often termed dissociated sensory paralysis, dissociated anæsthesia, or partial sensory paralysis.

4. *Muscular Sense.*—Under this term are included a number of different sensations: (1) sensation of passive movement; (2) the sense of position; (3) the sensation of weight and resistance.

(*a*) The first form may be tested by grasping the limb (or part) evenly all round, and making movements in various directions. Thus the movements of the great toe may be tested by the examiner holding its terminal phalanx between his thumb and index finger, making equal pressure with both, and slightly extending or flexing the toe. The patient should not contract any of the muscles of the toe during the

movement, but should allow the foot and toes to remain in a passive state. In the normal condition, the sensation of passive movements at the various joints is so delicate, that the movements through a very small angle are at once felt and their direction recognized. (Goldscheider estimates that, at the metatarso-phalangeal joint of the great toe, movement through an angle of 2° is appreciated : at the knee, movement through an angle of 0·5° to 0·7° is recognized.) According to Goldscheider the sensation is so delicate, that, at the majority of joints, movements which are only just visible to the examiner are felt by the patient. When the patient can only recognize movements which the examiner estimates to be to the extent of several degrees, then an affection of the sensation of passive movement is present.

(*b*) The sense of position is tested by placing a limb of the patient in a definite position when his eyes are closed. He is then asked to describe the position, or if the limb on the opposite side is unaffected, he is asked to place it in a similar position. In moving the limb it should be grasped evenly all round.

(*c*) The sense of resistance and weight may be tested by putting different weights in a sling placed over the limb (or portion thereof) which is to be tested. For the leg, a stocking with a small pocket attached has also been used, and for the hand cricket balls containing different amounts of lead in their interior.

In the normal condition a difference of one-tenth, i.e. the difference between 90 and 100 grammes, may be distinguished in the hand. In the leg the sensation is less delicate. In diseased conditions the results should be compared on the two sides, and also compared with those obtained in the normal state.

5. *Pressure Sense : Deep Sensibility.*—This is the sensation produced by pressure on the deep part of the limbs or trunk (by pressure on muscles, tendons, periosteum, fasciæ). The pressure may be made by the finger or by an instrument—the algometer (used by Head). The fibres subserving this form of sensation run mainly with the motor nerves, and are not destroyed by division of all the sensory nerves to the skin (Head).

[The views of Head, Rivers and Sherren on the three systems of fibres in the afferent peripheral nerves is mentioned on p. 103.]

6. *The Vibrating Sensation.*—When the foot of a vibrating tuning fork is placed over subcutaneous bony prominences or surfaces, in many parts of the body, a peculiar vibrating sensation is felt. To this sensation the names of vibrating sensation, or feeling, bone sensibility, and pallæsthesia have been given. Egger, in France, especially directed attention to the sensation in 1899.[1] Points which are suitable for testing are the following : styloid process of ulna, malleoli, inner surface of tibia, the sternum, the palms of hands, soles of feet, nails of fingers

[1] In the *British Medical Journal*, July 20, 1907, the author has given the results of observations in a number of cases.

and big toes. A large tuning fork is necessary for testing. The following are my conclusions respecting the vibrating sensation, which are based on a large number of observations.

(*a*) The vibrating sensation is a delicate test for detecting slight impairment of sensation. The vibrating sensation may be lost when other forms of sensation (to tactile impression, pain, and temperature) are felt quite well, or are only very slightly impaired. This is sometimes the case in early tabes, in slight peripheral neuritis, and often in diabetes mellitus.

(*b*) In diabetes mellitus, the vibrating sensation may be lost on the feet, or feet and legs, when there are no other nervous symptoms; but often the latter are present. In many cases of diabetes the nervous symptoms are chiefly (*a*) severe pains, tenderness and hyperæsthesia in the legs, (*b*) loss of the tendo Achillis jerks, (*c*) loss of the vibrating sensation.

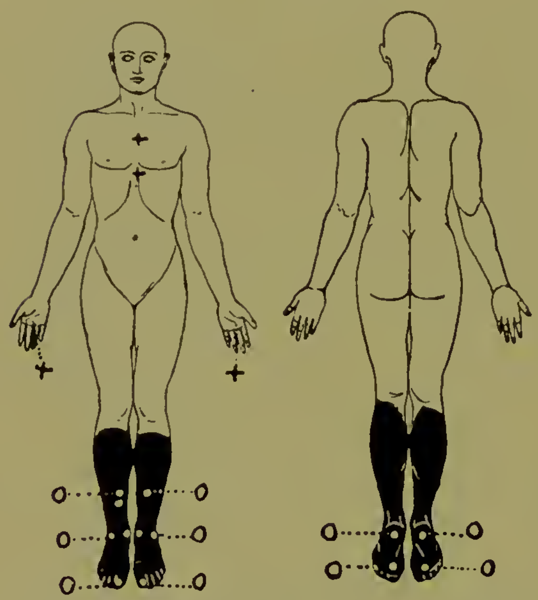


Fig. 50.—Diabetes Mellitus. Vibrating feeling lost at parts shaded and marked O; felt at parts marked +.

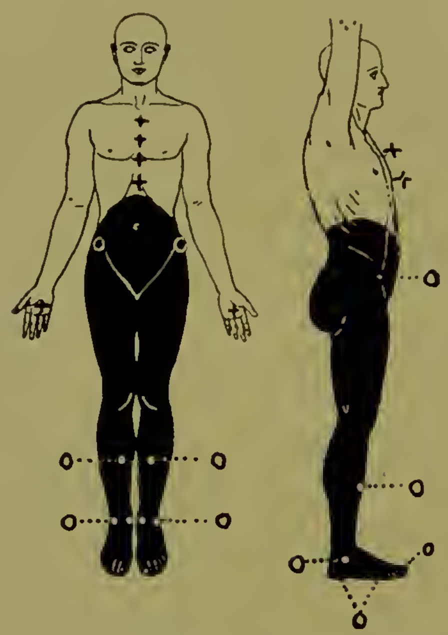


Fig. 51. — Spinal caries. Vibrating feeling lost in shaded area at parts marked O: felt at parts marked +

(*c*) In diseases strictly limited to the motor structures, the vibrating sensation is not lost even at an advanced period of the disease. (Thus it is not lost at an advanced period of amyotrophic lateral sclerosis.) In any case in which the disease appears to be one causing lesion only of the motor parts of the nervous system, if it be found that the vibrating sensation is lost, this fact indicates that the disease is affecting also sensory structures, and thus it may be of much diagnostic value.

In cases of paraplegia from spinal caries, and occasionally in spinal syphilis, the loss of the vibrating feeling may be the only objective symptom of affection of sensation at an early stage of the disease.

(*d*) In hemi-anæsthesia, if the vibrating feeling is not felt when the foot

of the tuning fork is placed on the edge of the sternum on the side of the tactile anæsthesia (at A in Fig. 52), but felt when placed at a corresponding point on the other side (B in Fig. 52) the case is one of hysterical or functional hemi-anæsthesia or of malingering.[1] If the vibrating sensation is felt distinctly when the foot of the vibrating tuning fork is placed on the edge of the sternum on the side of the tactile anæthesia, the hemi-anæsthesia may be due to organic lesion *or* to hysteria or neurosis.

7. In addition to the ordinary forms of sensation there is another—the *stereognostic sense*—by which the patient recognises the nature of objects placed in the hand, when the eyes are closed.

This is no isolated sensation; it represents the conclusion formed from various sensations — sense of movement, position, pressure sense, touch sensation, etc. Hence this sensation is impaired or lost through disturbance of various forms of sensation, tactile, muscular, etc. It may be lost when the sense of position only is affected. Also, when all forms of sensation are normal (for touch, pain, temperature, etc.) the sense may be lost through disturbance of the associative activity of the brain. The stereognostic sense may, therefore, be impaired through disease of the peripheral nerves and spinal cord causing affection of the separate forms of sensation on which the judgments of the nature of objects are formed. This is the cause of the loss of the stereognostic sense in rare cases of tabes. In cerebral affections, when the separate forms of sensation are normal, loss of the stereognostic sense (astereognosis or touch paralysis), is often due to cortical disease at the region of the inferior parietal lobule. It has been observed in softening, tumour, traumatic lesion, etc., of this region, and in cases of hemiplegia dating from birth or a very early period of life. In the last-mentioned condition other forms of sensation have been found normal, and the astereognosis may be really due to the stereognostic sense never having been developed. The loss of the stereognostic sense, through

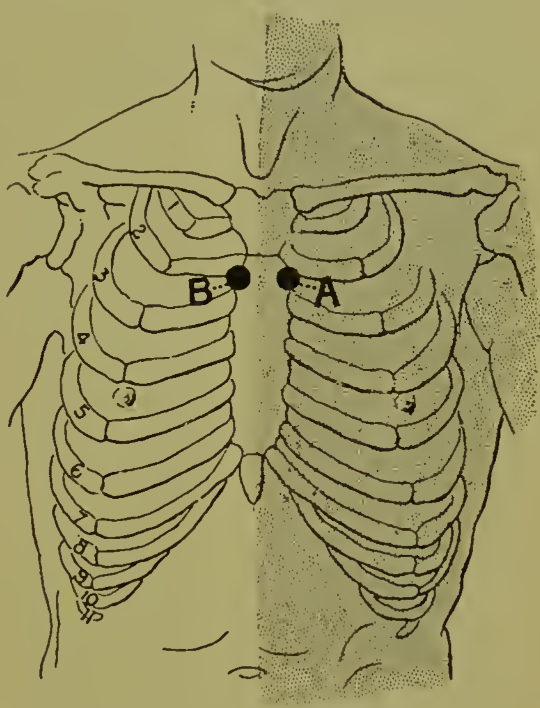


Fig. 52.—Hemi-anæsthesia. Anæsthetic side shaded.
In functional hemi-anæsthesia vibrating sensations *may* be lost at A, but felt at B.

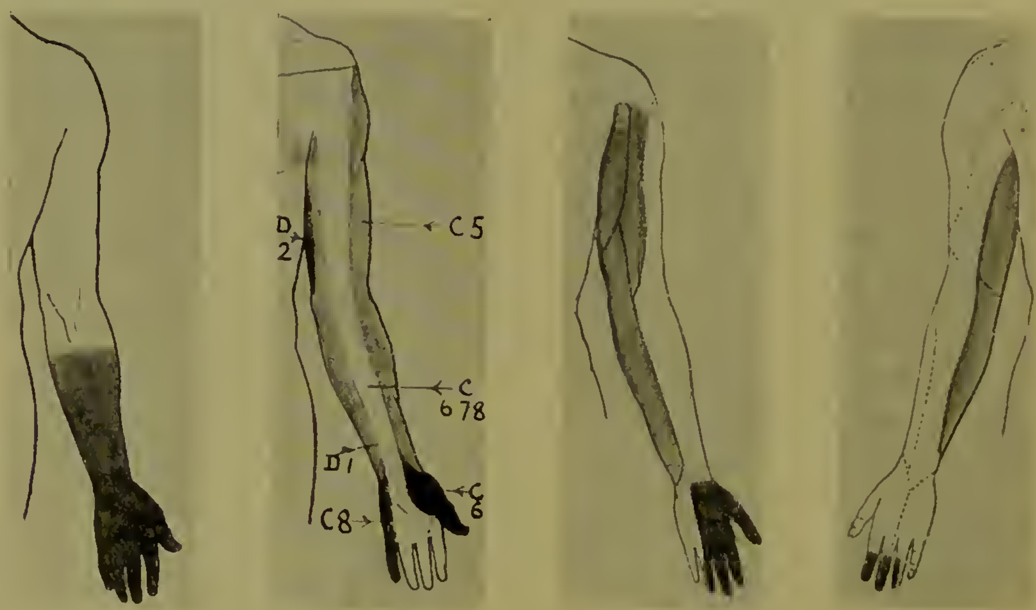
[1] The vibrations of the tuning fork are conducted readily all over the sternum. In organic hemi-anæsthesia, if the foot of the vibrating tuning fork be placed on the edge of the sternum in the anæsthetic half, the vibrations are conducted across the sternum and felt on the side which is not anæsthetic.

cortical lesion, is probably a condition analagous to word blindness and word deafness—really a condition of tactile aphasia.

II. Subjective Sensations.—The most important are pain and paræsthesia. Pain is a prominent symptom in spinal meningitis, in meningeal tumour, in caries and tumour of the vertebræ. Shooting pains in the limbs and girdle pains are prominent symptoms in tabes.

Paræsthesia, numbness, tingling, pruritus, pins and needles sensations occur in the early stages of many spinal diseases.

Sensation in the Muscles. In the normal condition severe pinching of the calf muscles causes muscular pain ; but in tabes often no pain



I. II. III. IV.

FIG. 53.—Anæsthesia in the Arm.

I. Anæsthesia (partial) from cerebral lesion.
II. Spinal lesion. Anæsthesia caused by lesion of spinal segments or nerve roots in shaded bands, the margins of which run more or less parallel with the long axis of the limb. D = dorsal. C = cervical segments. The numbers indicate the segments.
III. and IV. Anæsthesia caused by a lesion of a peripheral nerve (median) III. = Anterior surface of arm. IV. = Posterior surface.

is produced thereby, the sensitiveness of the muscles being greatly diminished or lost.

In tabes pinching of the arm muscles may cause pain, when the calf muscles are not sensitive. A strong faradic current causes pain when applied to normal muscles, but in tabes this painful sensation is often lost.

In peripheral neuritis the muscles are usually abnormally sensitive and painful on pressure (muscular hyperæsthesia or hyperalgesia). The same condition is also observed in many cases of diabetes mellitus.

General remarks on Sensory Disturbances.—Anæsthesia in rare instances may affect the whole of the body, with the exception of small

areas around the nose, the nipple and the genital organs. Such cases are usually due to hysteria.

Anæsthesia may affect the whole of one side of the body—(face, arm, trunk and leg)—hemi-anæsthesia : in such cases the cause is hysteria or an organic cerebral disease (usually affecting the sensory fibres of the internal capsule). A unilateral lesion of the spinal cord may cause anæsthesia below the level of the disease on the side opposite to the lesion.

In cases of cerebral disease, chiefly cortical lesions, and also in hysteria, anæsthesia may affect one portion of a limb—the hand or lower part of the arm. In cerebral disease, when there is loss of sensation in a portion of a limb only, the upper limit of the anæsthesia forms a line across the limb, more or less at right angles to the long axis of the limb. This may also occur in hysteria.

In spinal disease, when there is anæsthesia of only a portion of a limb the boundary line of the anæsthesia usually (but not invariably) runs more or less parallel with the long axis of the limb (*see* Fig. 53).

Anæsthesia when caused by the lesion of a peripheral nerve is limited to the distribution of that nerve.

A lesion of a spinal posterior nerve root causes anæsthesia in the distribution of that root. This area differs from that supplied by a peripheral nerve. Zones and bands of anæsthesia around the chest are often caused by root lesions in tabes.

In some spinal diseases there is no loss of any form of sensation—as in acute and chronic anterior poliomyelitis, amyotrophic lateral sclerosis, progressive muscular atrophy, primary lateral sclerosis.

Total anæsthesia (loss of all forms of sensation) below a spinal lesion is not very common. It indicates a complete transverse lesion of the cord and is a sign of grave prognostic significance.

Much more frequent than complete anæsthesia is partial anæsthesia. In some cases there is anæsthesia to one form of sensation, whilst other forms are preserved.

Analgesia and thermo-anæsthesia may occur when the disease affects the posterior horn of grey matter. This condition is common in syringomyelia, and in spinal hæmorrhage affecting the posterior grey matter ; it occurs in some forms of spinal syphilis, in many cases of Brown-Séquard's paralysis of syphilitic origin, and occasionally in tabes. I have observed the condition in acute poliomyelitis affecting both the anterior and posterior grey matter, and it has been noted in a few other diseases.

Sensation for temperature may be lost when sensation for pain and other forms of sensation are normal, as in some cases of spinal syphilis. Also in some cases of spinal syphilis there is loss of sensation to cold and perverted sensation (cold objects causing a sensation of warmth), whilst sensation for warmth and all other forms of sensation are normal.

Trophic Changes.

In the section on motor symptoms, a description has been already given of the atrophy of muscles which occurs, when there is a lesion of the lower motor neuron.

Trophic changes in connexion with the joints occur often in tabes (Charcot's joint disease), and occasionally in syringo-myelia. In tabetic joint disease, at first serous effusion occurs unassociated with pain or fever; later deformities arise—dislocations and subdislocations, erosion of the articular surfaces of the bones, and destruction of ligaments (*see* description, p. 312).

In infantile (spinal) paralysis the growth of the bones of the affected limbs is retarded.

Spontaneous fractures may occur in tabes, owing to the abnormal fragility of the bone. The cortical substance is thinner, the medullary spaces and Haversian canals are dilated, the whole bone is more porous, and a process of slow decalcification occurs. This altered condition of the bones causes a slight difference in the density of the shadow in the X-ray photographs.

Falling out of the teeth, with atrophy of the alveolar process, and falling off of the nails may occur in advanced cases of tabes.

Diseases of the spinal cord have usually no direct action on the general nutrition; but it may be mentioned, that *general wasting* is often noted in cases of tabes.

Bed sores, usually over the sacrum, with sloughing and deep ulceration, occur in some spinal diseases, especially in myelitis and other lesions of the lumbar region of the cord.

Perforating ulcers, chiefly on the feet, may occur in tabes, syringo-myelia, and diabetes mellitus (*see* p. 311).

Urticarial rashes and ecchymoses may follow the shooting pains of tabes. They occasionally occur in syringomyelia.

Herpes zoster occasionally follows or accompanies neuralgic pains, and is localised to the distribution of a sensory spinal nerve root. (In the majority of cases it is due to an affection of the spinal ganglion.) Herpes zoster is occasionally observed in tabes, myelitis, syringomyelia, and in caries and other diseases of the vertebræ.

Pemphigus has been observed in rare cases of syringomyelia.

Pigmentation of the skin, vitiligo, or a patch of grey hair may develop after neuralgic pain.

Leucoderma of the neck is often due to previous spyhilis.

A slight œdema occasionally occurs in limbs which have been paralysed for a long period.

In rare cases of tabes and syringomyelia, occasionally in myelitis and neuritis, and after traumatic nerve lesions, the skin of the affected part becomes atrophied and dry, and the surface smooth and shining (atrophy of skin, or glossy skin).

Ichthyosis is another rare condition occasionally met with in myelitis

and neuritis. In both conditions, glossy skin and ichthyosis, the sweat secretion is diminished or arrested. Scleroderma has been described occasionally in spinal diseases.

Vaso-motor disturbances are occasionally associated with spinal diseases. Vaso-motor paralysis is common in the paralysed limb in acute anterior poliomyelitis of the infant, and cyanosis and coldness of the limb are the result. In some spinal cases there is flushing of the skin, with or without increased perspiration. Abnormal vaso-motor reflex excitability occurs in some neurotic individuals, so that a stroke of the finger nail on the skin produces a raised red line, which persists for some time—(dermographia).

Paralysis of the Bladder and Rectum.

Loss of control of the bladder and rectum may be due (1) to disease of the centres for the reflex action of these organs ; or (2) to disease of the tracts above these centres, the voluntary (cerebral) control being thereby cut off. As regards the rectum, in the latter case, the sphincter ani still acts reflexly ; and if the finger be introduced into the rectum there is first a relaxation and then a firm tonic contraction of the sphincter, showing that the centre is intact. But if the centre be destroyed, or the nerve fibres from it interrupted, no tonic contraction of the sphincter ani can be felt : " there is complete and persistent relaxation " (Sir W. Gowers). When the centre for the sphincter is destroyed paralysis of that muscle and incontinence of fæces occur. In lesions above the centre the influence of the will is cut off, and reflex involuntary evacuations of the rectum occur.

As regards the bladder, if the centres at the lower end of the cord be destroyed, the sphincter fails to act, and urine dribbles away as soon as it enters the bladder.

But if the disease causing paralysis of the bladder be above the reflex centres, the cerebral influence is cut off and the bladder may act automatically. In some cases, after a certain amount of urine has collected in the bladder, the muscle wall contracts, the sphincter relaxes, and urine is passed—intermittent incontinence. In other cases the muscular wall of the bladder fails to contract and retention of urine occurs. When the bladder remains over-distended, the pressure becomes so great, that in time urine is forced through the sphincter and dribbling occurs from the distended bladder—overflow incontinence. The *total* destruction of the transverse section of the cord at any level is said to abolish the reflex activity (power of reflex evacuation) of both bladder and rectum.

When the bladder is paralysed the urine often becomes alkaline or ammoniacal, the mucous membrane of the bladder is very liable to become inflamed (cystitis) ; the kidneys are liable to be affected secondarily (pyelitis, pyelo-nephritis, and abscess) and these complications are common causes of death in spinal diseases, through septic infection.

Ocular Symptoms.

The Oculo-Pupillary Phenomenon.—This is a useful localising sign. It consists in unilateral contraction of the pupil, with diminution of the palpebral fissure and slight sinking in of the eyeball. The cause is paralysis of certain fibres of the cervical sympathetic nerve. These fibres arise in the cilio-spinal centre at the upper part of the cord; they pass through the anterior roots of the first dorsal nerve, and then through the rami communicantes to the cervical sympathetic nerve. According to some physiologists fibres also pass through the second dorsal nerve, according to others through the eighth cervical. From the cervical sympathetic the fibres pass to the dilator fibres of the pupil, and, according to some authors, to the muscle fibres of Müller, at the back of the orbit. Claude Bernard found that the ocular symptoms just described were produced by section of the anterior roots of the first and second dorsal nerves. Mlle. Klumpke produced them (without vaso-motor symptoms) by section of the eighth cervical and first dorsal nerve roots in dogs.

Hutchinson, many years ago, pointed out the localising value of the symptom and suggested the explanation. Through clinical observations in cases of paralysis of the lower part of the brachial plexus, and through experiments on animals, Hutchinson's view has been confirmed.

In man these oculo-pupillary symptoms are often unilateral and are caused by lesion of the first dorsal and eighth cervical anterior nerve roots. In these cases there is paralysis of the brachial plexus of the lower arm type, and anæsthesia is present along the inner (ulnar) side of arm, forearm and hand; there may be also absence of sweat on the face and upper part of the thorax on the affected side.

In lesions of the cord the symptoms are less constant.

In paralysis of the cervical sympathetic the cutaneous vessels of the face and scalp may be dilated from vaso-motor paralysis; this condition is succeeded by contraction of the vessels.

If the nerve fibres (already described) are not destroyed, but simply irritated, as in cases of vertebral caries, the pupil is dilated, the palpebral fissure wider and the eye-ball a little more prominent: also there is pallor and sweating of the affected side of the face. When vaso-motor symptoms are present on the face the lesion extends further down the dorsal region (in the region of the upper dorsal vertebræ).

In man, lesions giving rise to irritation of the sympathetic very often produce paralysis of it at a later stage.

Lesions of the nerve roots causing sympathetic paralysis must be close to the cord—before the rami communicantes are given off.

In paralysis of the sympathetic oculo-pupillary fibres, the pupil does not dilate on shading the eye; it does not dilate when cocaine is dropped into the eye; also the cilio-spinal reflex may be abolished on the side of the lesion, i.e. the pupil does not dilate when the skin of the neck is firmly pinched. But the pupils contract to light and on

convergence. Symptoms of irritation of the cervical sympathetic can be caused by dropping a little cocaine solution into the eye.

Traumatic lesions of the cervical sympathetic in the neck, or compression by aneurism, tumour or other lesions in the thorax, may produce symptoms similar to those just described.

Pupillary Reflexes.—A reflex of great diagnostic value is the reflex contraction of the pupil on exposure to light, after it has been shaded. In the normal condition the pupil contracts both on the side stimulated by the light and on the opposite side (consensual reflex). In most cases of tabes, in some cases of general paralysis of the insane, and in some syphilitic individuals, the pupils do not contract to light, but contract when the eyes accommodate for near objects (Argyll-Robertson pupil, *see* p. 308).

In cerebro-spinal syphilis the pupillary reflexes both to light and accommodation, are often lost.

In tabes often the pupils are very small (myosis); in some diseases the pupils are dilated (mydriasis); also they may be unequal, as in general paralysis of the insane and other affections.

Paralysis of the third cranial nerve or irritation of the cervical sympathetic will cause dilatation of the pupil on the affected side. In paralysis of the cervical sympathetic, as just mentioned, the pupil is contracted on the affected side.

Ophthalmoscopic examination is not so helpful in the diagnosis of diseases of the spinal cord, as in cerebral affections; but in several diseases it furnishes valuable indications. Optic atrophy is a valuable sign in tabes and disseminated sclerosis.

In syphilitic cord diseases, disseminated choroiditis or syphilitic retinitis is an occasional symptom of value in the diagnosis. Optic neuritis has been detected in many cases of disseminated myelitis (*see* p. 133).

Occasionally in lesions of the cervical region of the spinal cord (tumour, myelitis, compression myelitis, etc.) optic neuritis has been detected, though pathological examination has not revealed any intracranial tumour or meningitis. Headache and vomiting may be singly or together associated with the optic neuritis of such local lesions of the cervical cord (*see* paper by Taylor J. and Collier, J. *Brain*, p. 532, 1901).

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SECTION V

ELECTRICAL EXAMINATION

In many diseases of the cord electrical examination is of diagnostic value. In the normal condition stimulation of a motor nerve of a limb, both by faradism and by galvanism, causes a contraction of the muscle it supplies. Direct stimulation of a muscle (at its motor point), both by faradism and galvanism, causes a very sharp contraction; and the kathodal closing current causes a more powerful contraction than the anodal closing current. (Using the letters KCC for Kathodal closing contraction, and ACC for Anodal closing contraction, this relation is expressed as KCC > ACC.) Te in the following description indicates tetanus of the muscle, and OC indicates opening contraction.

In tabular form the normal reactions are as follows :—

Nerve.—Reaction to weak faradic and galvanic currents.

Muscle.—Reaction to weak faradic current.

Reaction to galvanic current. Contraction of muscle instantaneous. KCC > ACC.

With galvanism the contractions obtained in health are as follows :—

Weak current.—KCC only.

Stronger.—KCC, ACC, AOC.

(Usually ACC > AOC.)

Stronger.—KCTe, ACC, AOC.

Very strong.—KCTe, ACTe, AOC, KOC.

Only an exceedingly strong current produces AOTe ; usually it is not obtained in health.

Electrical Reactions in Diseases.—In some diseases there is a simple diminution of the electrical excitability without other change, as in the atrophy of muscles from disuse, in the atrophy due to hysterical paralysis, and in primary muscular diseases. Simple increase of electrical excitability is rare. It is a marked symptom in tetany ; the KCC is obtained by a very weak current, and by a relatively weak current the contractions become tetanic, KCTe ; also AOTe, and even KOTe may be obtained.

The change which is of the most importance in diagnosis is the *reaction of degeneration*, an abnormal condition of electrical excitability found in certain diseases. In the *complete* reaction of degeneration the condition is as follows :—

1. The excitability of the *nerve*, both to the *faradic* and *galvanic* current, is lost.

2. The excitability of the *muscle* to *faradism* is lost.

3. The excitability of the *muscle* to *galvanism* is retained :

(*a*) It is increased at first, afterwards diminished.

(*b*) The muscular contraction is *sluggish.*

(*c*) There is often a qualitative change in the excitability. ACC is more powerful than KCC ; or ACC and KCC are equal ; or sometimes the normal condition is found, KCC being greater than ACC. (i.e., ACC $>$ KCC ; or ACC $=$ KCC ; or KCC $>$ ACC).

The galvanic excitability of the muscles showing the reaction of degeneration is only increased at first ; this excitability then gradually diminishes ; and after several years, if the paralysis should continue, a strong galvanic current only gives a very slight contraction—so slight that the contraction causes no movement of the limb or of its segment, but if the attention be directed to the muscle itself a slight wave of contraction can be seen.

It is remembered that ACC is not always greater than KCC in muscles which otherwise present the changes of the reaction of degeneration ; sometimes ACC $=$ KCC, and sometimes KCC $>$ ACC, even when the other changes of the reaction of degeneration are present. Also Oppenheim points out that occasionally in a normal muscle the ACC may be greater than the KCC. In the normal condition, the contraction of the muscle to galvanism is sharp and short—lightning contraction. In muscles which show the reaction of degeneration, the contraction is sluggish and worm-like. The *sluggish contraction* of the muscle is the *most* important galvanic change of the reaction of degeneration.

Besides the complete form just described, not infrequently a *partial* reaction of degeneration is met with. In this partial form the condition is as follows :—

1. The excitability of the nerve to faradism and galvanism is diminished but not lost.

2. The excitability of the muscles to faradism is diminished or lost.

3. The excitability of the muscle to galvanism is *sluggish* and ACC is greater or equal to KCC. Between the partial and complete form of the reaction of degeneration, there are intermediate forms which are of no practical importance.

It may be stated that, with a few exceptions, the reaction of degeneration indicates a lesion of the lower motor neuron ; and that lesions of the upper motor neuron and primary lesions of the muscles do not cause this reaction.

Limits of space do not permit a description of the general methods of electrical examination in a work devoted especially to disease of the spinal cord.

In the *Manchester Medical Students' Gazette* of January 1903, the author has given a short sketch of the methods of electrical diagnosis. For further information the works of Dr. Lewis Jones (*Medical Electricity*, London,

1906), and of T. Cohn (*Leitfaden der Elektrodiagnostik und Elektrotherapie*, Berlin, 1906), may be recommended.

In the following table the typical changes in the reaction of degeneration are compared with the normal condition.

Stimulation.		Normal.	"Reaction of Degeneration."
Of *Nerve*	Faradism	Contraction (to weak current)	No contraction.
	Galvanism	Contraction (to weak current)	No contraction.
Of *Muscle*	Faradism	Contraction (to weak current)	No contraction.
	Galvanism	Contraction	Contraction.
	(*a*) Jerk of Muscle	Sharp: lightning	*Sluggish: prolonged.*
	(*b*) Polar excitability	Kathodal closing contraction the greater —KCC > ACC	Anodal closing contraction equal to or greater than the kathodal—ACC = KCC, or ACC > KCC.

Examination with the X Rays.

It has been shown by many observers that the X Ray examination is of value in diagnosis in diseases of the vertebræ (caries, tumour, fracture and dislocation) and for the detection of bullets in the vertebræ or neighbourhood of the spinal cord and cauda equina. Also in Charcot's joint disease and other affections of joints and bones occurring in affections of the spinal cord, the X Ray examination is often of service in demonstrating the nature of the changes.

For further details, *see* article by Leyden and Grunmach, *Archiv. f. Psychiatrie und Nervenkrankheiten* Bd. 37, 1903. (Abstracted by the author in the *Medical Chronicle*, January 1903.)

Lumbar Puncture.

The examination of the cerebro-spinal fluid is of diagnostic value in several diseases. The fluid is withdrawn from the spinal subarachnoid space by means of a fine exploring trocar and cannula, introduced in the lumbar region of the spine. This procedure is known as lumbar puncture. It was first introduced by Prof. Quincke in 1891; and Widal, Sicard, Ravout and many others have demonstrated its diagnostic value. In rare cases it has been of service in treatment.

Method of Obtaining the Puncture Fluid.—The patient is placed on the left side, in the horizontal position, with the vertebral column bent forwards as much as possible. The interval between the 3rd and 4th or the 4th and 5th lumbar vertebral spines is determined. A line drawn across the patient's back connecting the uppermost part of each iliac crest will be at right angles to the vertebral column, and will cross the middle line between the 3rd and 4th lumbar spines. The puncture may be made at this point (between the laminæ of the 3rd and 4th

lumbar vertebræ) or between the 4th and 5th laminæ. The spinal cord ends at the upper border of the second lumbar vertebra. Below that point, within the spinal dura mater, are the nerve roots of the cauda equina, surrounded by cerebro-spinal fluid. Hence from this region the cerebro-spinal fluid can be withdrawn without injuring the spinal cord.

The skin of the back in the lumbar region, around the point of puncture, is sterilised by a disinfecting solution. An anæsthetic is usually not necessary, but if the pain of the puncture be feared, the skin may be made anæsthetic by the action of ethyl chloride spray. In children the puncture is made in the middle line, in adults one centimetre (or nearly half an inch) to one side of the middle line. A special fine trocar and cannula, or the needle of an exploring syringe, may be used. If the latter be used the piston of the syringe should be removed. The needle (or trocar) is pushed slightly upward (i.e. directed slightly towards the upper end of the spine) and slightly towards the middle line (in adults). The needle, which has been sterilised by boiling, should be at least 8 cm. (a little over 3 inches) long. The needle is pushed forwards, in the direction mentioned, until the resistance is felt to cease, owing to its passage through the ligamentum subflavum and spinal dura mater into the subarachnoid space. If the point of the needle be obstructed by bone it should be withdrawn. The needle should be introduced for about 2 cm. in the child, for about 4–6 cm. in the adult. For diagnostic purposes only a small amount of fluid should be allowed to flow away—1 to 2 c.c. The fluid should not be withdrawn by aspiration, it should be allowed to flow away slowly, drop by drop, and should be collected in a glass vessel. The pressure of the fluid may be measured by attaching to the puncture needle a manometer, or a glass tube held vertically. The fluid rises in this glass tube, the height it reaches being an indication of the pressure of the cerebro-spinal fluid. Normally the fluid pressure is equal to 125 m.m. of water (the lumen of the tube having a diameter of 1 m.m.).

When the pressure is normal 1–5 c.c. of fluid may be removed, but the fluid should not be allowed to flow after the pressure has sunk to 100 or 80 m.m. When the pressure is increased at first, the fluid should not be allowed to flow away after the pressure has sunk to 120 m.m. After the fluid has been collected the needle is withdrawn, and the skin puncture must be closed by celloidin or collodion. The patient should be kept in bed for twelve or twenty-four hours after the puncture. Frequently the operation is followed by headache.

The *normal* cerebro-spinal fluid is transparent and colourless, like water. The sp. gr. is 1006 to 1007, and the reaction alkaline. The fluid contains a trace of serum globulin and albumose, and a substance which reduces Fehling's solution (not sugar). It contains a very few lymphocytes and large endothelial cells, but both are very difficult to find. It does not contain micro-organisms.

In *pathological conditions* the examination for micro-organisms

and cells is sometimes of diagnostic value, and the physical and chemical changes are occasionally important. By accidental puncture of an arachnoid vein blood is, in rare cases, mixed with the fluid, but in a few seconds the fluid usually becomes clearer. In fracture of the skull, in subarachnoid hæmorrhage (spinal or cerebral), in intra-ventricular hæmorrhages, and in injuries to the spinal cord blood may be mixed with the puncture fluid. In these cases the fluid remains yellow, even after the use of the centrifugal apparatus, and the fluid is equally tinged with blood from first to last. But when blood is mixed with the puncture fluid accidentally, the first few drops are most tinged, and after the use of the centrifugal apparatus, the blood corpuscles fall to the bottom of the tube, and the superjacent fluid is clear. In cerebral hæmorrhage, and in cerebral meningeal hæmorrhage, the puncture fluid may be tinged yellow, whilst in cerebral thrombosis it is not coloured. In severe jaundice the puncture fluid may be yellow. In many cases of meningitis the fluid is turbid, and occasionally purulent. On standing a fibrin clot may form at the bottom of the glass in tuberculous, syphilitic, and serous meningitis, and in sarcoma of the meninges.

The *pressure* of the fluid is increased in intra-cranial tumours, in blood extravasation in the brain or membranes, in serous or purulent extravasation in the ventricles of the brain or subarachnoid space. The pressure may rise up to 300 m.m., seldom to 700 m.m. of water, the normal pressure being 125 m.m.

Normally, and in cases of hydrocephalus, the amount of albumen is 0·2 to 0·5 per cent. ; often it is only a trace. The proteid is chiefly serum globulin, and is precipitated by an equal volume of saturated ammonium sulphate solution. A small amount of serum albumen is also present. Both are precipitated by acidification and boiling of the fluid. In pathological conditions (as in purulent meningitis) the albumen is increased—up to 2 to 8 per cent. The amount of albumen may be estimated by a modification of Esbach's albuminometer.

Urea has been found in the cerebro-spinal fluid in uræmia. In diabetic coma the puncture fluid has given the same reaction as diabetic blood when the author's methylene blue test has been employed.[1]

Bacteriological examination of the fluid has revealed various micro-organisms and parasites. Cover-glass preparations of the fluid may be stained for the organism, or cultivations may be made from the puncture fluid. In suspected tubercular meningitis the fluid has been injected into guinea-pigs, and tubercular disease thus produced. The following organisms have been detected in various diseases : the tubercle bacillus (tubercular meningitis), the meningococcus intracellularis (epidemic cerebro-spinal meningitis) ; staphylococci, streptococci, pneumococci (in other forms of meningitis) ; trypanosomata (in sleeping sickness).

[1] See author's article in the *British Medical Journal*, September 19, 1896, and the *Lancet*, August 4, 1900, and R. Müller's paper, *Münchener med. Woch.*, No. 25, 1899.

Character of the Cells.—In some pathological conditions the cells in the fluid are greatly increased. As already mentioned, normally cells are found with difficulty, the númber being from 0·5 to 2 in one cubic millimetre of fluid. In general paralysis of the insane there may be as many as 60 cells in 1 c.mm. ; in tabes and in meningitis of various forms the cells are often greatly increased in number.

A useful method of examination for cells is to centrifuge 5 c.c. of the puncture fluid for five minutes, to empty the tube, turning it upside down, and to scrape the bottom with a fine capillary pipette. The sediment is transferred to a slide, fixed by heat, stained by Jenner's methylene blue and eosin stain, and mounted in Canada balsam (Widal, Purves Stewart).

Normally the cerebro-spinal fluid, when examined in this manner, contains no polymorpho-nuclear leucocytes, and only an occasional mono-nuclear lymphocyte, with now and then a few endothelial cells. Examination of the centifuged deposit with a magnification of 400 diameters (in the manner just described) should not show more than two or three lymphocytes in the microscopic field (P. Stewart). Sometimes no cells can be seen. In some diseases the cells are greatly increased in number. In acute meningitis, especially in suppurative forms, there is usually a marked *leucocytosis* of the puncture fluid, in which the cells are mostly polynuclear (or polymorphs). When recovery commences the polynuclear leucocytes diminish, and are replaced by lymphocytes. In tubercular and syphilitic inflammation, and in tabes and general paralysis of the insane, the cells are much increased in number and are *lymphocytes*—monomorphic cells. This is the general rule, but exceptions occasionally occur.

These changes in the cells of the fluid, and the detection of the tubercle bacillus, of the meningococcus intracellularis, and other organisms are of diagnostic value.

(The character of the puncture fluid in tabes, and in the various forms of meningitis, will be described in the chapters devoted to these diseases.)

Dangers of Lumbar Puncture.—Lumbar puncture has occasionally been followed by dangerous symptoms, and Gumprecht has collected records of fifteen cases in which death occurred directly after the puncture. In many of these cases the patient had suffered from cerebral tumour, and often very large quantities of fluid had been withdrawn.

In order to avoid bad results it is important that the fluid should not be aspirated, but should be allowed to flow away spontaneously and slowly. Only 1 or 2 c.c. should be withdrawn in most cases, and the patient should be kept lying down for three or four hours. There is usually slight headache after the puncture.

REFERENCES.

Further details as to the therapeutic and diagnostic value of lumbar puncture and references to the literature of the subject are given in a review by the writer in the *Medical Chronicle*, March, 1907.

SECTION VI

ON THE DIAGNOSIS AND LOCALISATION OF DISEASES OF THE SPINAL CORD

The diagnosis of the diseases of the spinal cord should be based on the results of a careful systematic examination of the patient, and "rapid diagnoses" are very liable to lead to mistakes.

The history of the case should be taken, a general examination of the patient made, and the condition of the nervous system investigated. The motor and sensory symptoms, the condition of the reflexes and the gait should be noted, and inquiries should be made respecting bladder and rectal symptoms. In *all* cases of paraplegia the *spine* should be examined for signs of caries, and the condition of the *bladder* should be determined by palpation of the lower part of the abdomen. An ophthalmoscopic examination is often necessary; in the *majority* of spinal affections it is desirable. Electrical examination is sometimes of diagnostic value.

From the extent and nature of the symptoms we form an opinion as to the seat of the disease, i.e. we *localise* the lesion, or form an *anatomical* diagnosis. From the mode of onset, the history and other features of the case we diagnose the *nature* of the lesion, i.e. we make a *pathological* diagnosis. We are then in a position to form a broad conception as to the disease of the cord in the case we are investigating.

Anatomical Diagnosis (Spinal Localisation).

Suggestive papers respecting spinal localisation were published many years ago by a former professor of medicine at the Owens College —the late Dr. J. Ross; and the whole subject has been very thoroughly worked out by Mr. William Thorburn, of Manchester, by Allen Starr, Edinger, Kocher, Sherrington, J. Mackenzie, Head and many other observers.

(*a*) **Spinal Localisation in the Vertical Direction.**—At definite intervals pairs of nerve roots (anterior motor and posterior sensory) are given off from the spinal cord on each side; and thus the spinal cord may be divided into segments corresponding to the origin of the nerve roots. Clinical and pathological observations, and experiments on animals, have shown that definite functions are localised to the various spinal segments and nerve roots.

Motor Localisation.—Some muscles are supplied by the motor nerve cells of *one* spinal segment and the motor fibres of *one* nerve root (intercostals and a few muscles of the limbs). But most muscles are supplied from more than one segment and receive usually nerve fibres from more than one motor nerve root. Most muscles of the limbs receive nerve fibres from three spinal segments (Sherrington).

Probably the motor fibres of a muscle spring from one main nucleus and several accessory nuclei. Moreover in one segment of the cord the nuclei of several muscles are situated.

According to Sherrington "no focal lesion of the cord can damage, far less completely paralyse, a single muscle alone; it will tend to weaken a number, but it cannot paralyse one alone."

Thorburn states, however, the point of practical importance is that each muscle appears to have only one main root of supply—a root the injury or disease of which suffices to cause paralysis in the muscle.

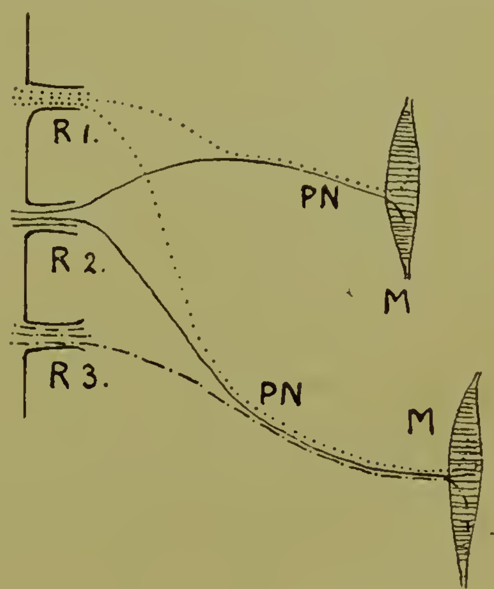

Fig. 54.—Diagram showing Nerve Supply of Muscles from two or three Spinal Nerve Roots. M = muscle. PN = peripheral nerve. R1, 2, 3 = Nerve roots leaving the Spinal Cord. The peripheral nerve supplying the upper muscle derives fibres from roots 1 and 2. The nerve supplying the lower muscle derives fibres from roots 1, 2, 3.

Probably the segment of origin of each motor root has motor functions corresponding more or less with those of the root.

In cases of spinal disease, in which there is motor paralysis, a careful examination is necessary to determine which muscles are paralysed and which are spared. Tables have been prepared by various writers, indicating the muscles paralysed in lesions of the various nerve roots. They enable us to localise the spinal lesion, as regards its vertical extent, in any case of spinal paralysis. The tables differ slightly in detail, but the following are the more important landmarks (taken chiefly from Mr. Thorburn's writings):—

Motor Localisation.

Nerve Root.	*Muscles.*
Cervical 4.	Diaphragm, scaleni, supra-spinatus, infra-spinatus (? rhomboids and teres minor).
C 5.	Biceps, deltoid, brachialis anticus, supinator longus, supinator brevis (? coraco-brachialis).
C 6.	Subscapularis, pronator teres, pronator quadratus, teres major, latissimus dorsi, pectoralis major (in part), triceps, serratus magnus (? pectoralis minor).
C 7.	Extensors of wrist (+ ? extensors of fingers).
C 8.	Flexors of fingers (+ ? flexors of wrist).

Nerve Root.		*Muscles.*
Dorsal	D 1.	Small muscles (intrinsic muscles) of hand, oculo-pupillary fibres.
	D 2.	
	D 3.	
	D 4.	
	D 5.	
	D 6.	Intercostal muscles.
	D 7.	
	D 8.	
	D 9.	
	D10.	Abdominal muscles.
	D11.	
	D12.	
Lumbar	L 1.	Quadratus lumborum.
	L 2.	Cremaster.
	L 3.	Sartorius, adductors of thigh, ileo-psoas.
	L 4.	Extensor quadriceps cruris, abductors of thighs.
	L 5.	Flexors of knee (hamstring muscles).
Sacral	S 1.	Calf muscles.
	S 2.	Glutei, peronei, anterior tibial muscles, intrinsic muscles of foot.
	S 3.	Muscles concerned in erection of the penis and ejaculation. Centres for erection and ejaculation.
Conus Terminalis Muscles of sexual organs, bladder, rectum.	S 4.	Levator ani and centres for sphincters of anus and bladder.

In addition to the localisation indicated in the table there are several points of interest which may be added.

A complete transverse lesion of the cord in the first four cervical segments is rapidly fatal through paralysis of respiration. If the lesion be unilateral or not completely transverse the duration of life is longer, and observations then indicate that the first three cervical segments supply the deep muscles of the neck.

According to many observers the deltoid, biceps, brachialis anticus and supinator longus are supplied by the 5th and 6th nerve roots.

The oculo-pupillary fibres of the sympathetic leave the cord by the 1st dorsal root, or by the 8th cervical and 1st dorsal.

Lesion at the first dorsal segment frequently causes oculo-pupillary symptoms : contracted pupil (myosis), retraction of the eye-ball and narrowing of the palpebral fissure.

Some observers believe that the first lumbar segment supplies the ileo-psoas.

A transverse lesion of the cord at the level of the 1st lumbar segment will cause paralysis of the legs and of the bladder and rectum ; at the level of the 2nd dorsal, the intercostal and abdominal muscles will be paralysed also ; at the level of the 1st dorsal, the small muscles of the hand will be paralysed in addition to all of the muscles just mentioned. A lesion just below the 5th cervical will paralyse all the arm muscles except the deltoid, biceps, brachialis anticus and supinator longus, as well as all of the muscles already mentioned ; at the level

of the 5th cervical the paralysis would extend to the four muscles last named. A lesion at the level of the 4th cervical segment would paralyse the diaphragm, and death would occur from asphyxia.

Localisation by Extent of Perspiration.—Sir V. Horsley has employed the injection of pilocarpine as a means of localising a cord lesion. The sweat glands of the skin supplied from the part of the cord above a transverse spinal lesion act well under the influence of pilocarpine, whilst those of the skin supplied from the cord below the level of the lesion do not, and thus at the junction of the two regions there is a line of small drops of sweat.

Horsley warns against the use of this test if there should be any tendency to bronchorrhœa.

Sensory Localisation.—The sensory distribution of the spinal nerve

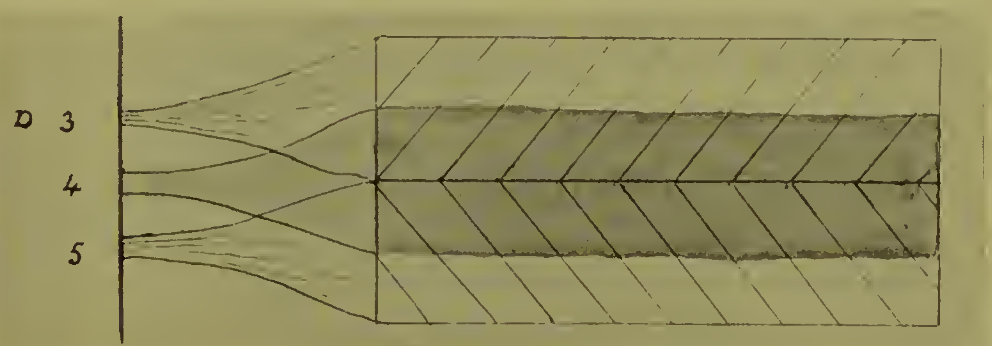

FIG. 55.—Diagram showing overlapping of Sensory Areas of Skin (modified after Sherrington). To the left are three sensory nerve roots entering the spinal cord. The skin area supplied by root 4 is shaded. The areas supplied by roots 3 and 5 are marked by oblique lines. Note that the upper half of the sensory distribution of root 4 is overlapped by the area of distribution of 3 ; its lower half by the area of distribution of 5.

roots has been determined by dissection, by experiments on animals, by clinical observations in cases of injury or disease of the spinal cord or its roots at various levels, by the distribution of the skin affection in herpes zoster, and by the areas of referred pain in visceral diseases.

Every posterior spinal nerve root supplies a definite skin area, but these areas overlap each other to some extent, at least so far as tactile sensation is concerned. "Any given area of skin seems to possess tactual sensory nerve fibres entering the cord via two spinal ganglia." "As in the musculature, so in the skin, the degree of overlapping of nerve root distribution for touch, as well as for pain, is greater at the distal end of the limb than elsewhere. In the skin of the hand and foot are regions innervated through three consecutive spinal ganglia, whereas in most parts of the limb, as in the trunk, the supply of any one area is by two consecutive roots only. The amount of overlap of the right with left across the median line varies in various parts ; in the tongue it is slight ; on the front of the chest it is considerable. The overlapping is greater for touch nerves than for pain nerves" (Sherrington).

According to Head, Sherrington's results are true for pressure sense

only, whilst for pain and temperature sensation there is a sharp limitation of the root areas.

In any definite skin area complete anæsthesia is only produced when all of the nerve roots supplying the area are affected. Hence in any definite area there is complete anæsthesia only when the highest of the nerve roots supplying it is affected. Moreover the spinal lesion is above the region of anæsthesia.

On the trunk the root districts surround the body in an obliquely

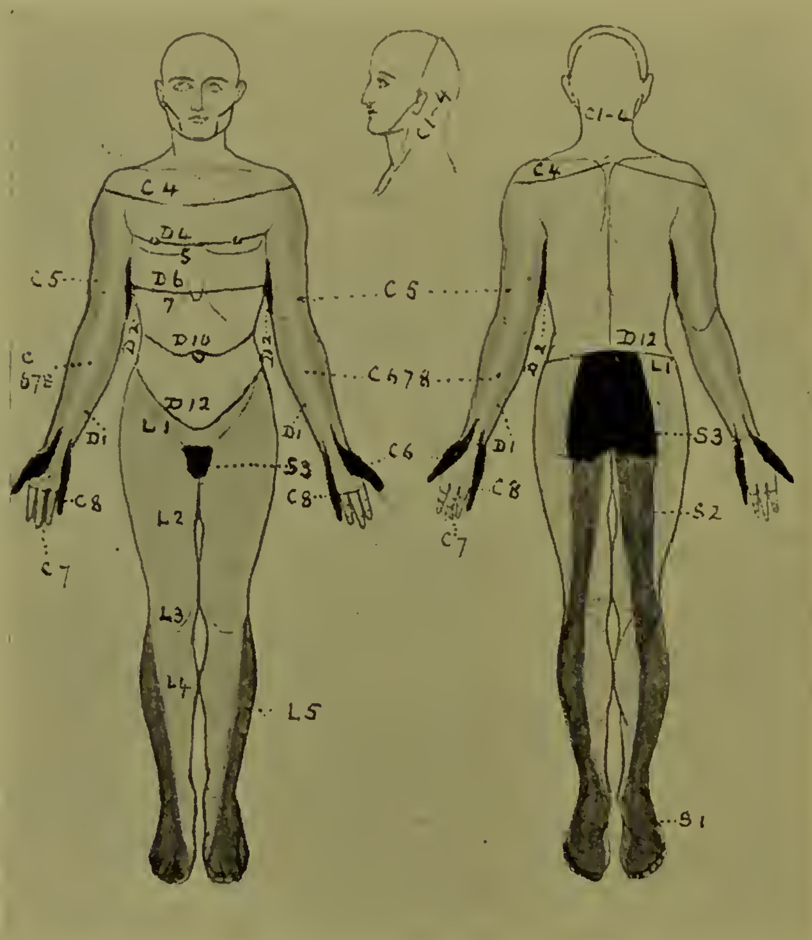

FIG. 56.—Diagram showing Sensory Areas supplied by various Spinal Segments and Nerve Roots.

C = Cervical. L = Lumbar.
D = Dorsal. S = Sacral segments.

transverse manner, in the limbs the areas of distribution of the sensory roots run more or less parallel to the long axis of the limb.

A lesion at the level of the 3rd sacral segment produces a saddle-shaped area of anæsthesia in the gluteal region, and the scrotum and penis (or the vulva) are also anæsthetic. At the level of the 2nd sacral segment there is, in addition, a strip of anæsthesia down the back of the thighs. When the lesion is at the 1st sacral segment, according to most observers, the anæsthesia extends down the leg to the sole of the foot, but there are considerable differences of opinion as to its exact limits. At the level of the 5th lumbar there

is the saddle-shaped area of anæsthesia in the gluteal region already mentioned, with a strip of anæsthesia down the back of each thigh, anæsthesia of the outer half of thé leg below the knee (anteriorly and posteriorly), and of the dorsum and sole of the foot; whilst the front of the thigh, and the inner side of the leg and foot are not anæsthetic (*see* Fig. 56). A lesion at the level of the 1st lumbar segment causes anæsthesia of the legs up to a line a short distance above the groin. The parts supplied by the 2nd, 3rd and 4th lumbar segments are roughly indicated in the diagram, but the exact limits of these regions are disputed (*see* Fig. 56). A lesion at the 10th dorsal segment causes anæsthesia up to the umbilicus; at the 6th and 7th dorsal, up to the ensiform cartilage; at the 4th and 5th dorsal, up to the nipples. The sensory distribution in the arm is shown in Fig. 56. A lesion at the 8th cervical segment produces a strip of anæsthesia down the inner side of the arm and hand and anæsthesia of the little finger. The face and anterior part of the scalp is supplied by the 5th cranial nerve; the posterior limit of its distribution is a line drawn from the crown of the head to the ear and chin. Below and behind this line is the area supplied by the first four cervical nerves (*see* Fig. 56).

(Fig. 56 shows the more important landmarks of spinal sensory localisation. The localisation in the arms is that given by W. Thorburn; the localisation in the legs and trunk is taken from Seiffer's diagram, which only indicates the best established sensory landmarks.)

The Distribution of the Rash in Herpes Zoster with respect to Spinal Localisation.—Over thirty years ago von Bärensprung attributed herpes zoster to a lesion of the ganglion of the posterior nerve root, and recorded changes—remains of blood extravasation, etc.,—found in the ganglion in this disease. Subsequently other cases were recorded and the subject has been carefully studied by Dr. J. Mackenzie, of Burnley, and others. Head and A. W. Campbell have recorded the pathological changes in twenty-one cases. They found that in herpes zoster the acute changes in the ganglion of the posterior root consist of—(1), an extremely acute inflammation with exudation of small round cells; (2) extravasation of blood; (3) destruction of ganglion cells and fibres; (4) inflammation of the sheath of the ganglion. If severe, the changes leave a scar in the affected part of the ganglion; if slight, the ganglion in time recovers its normal appearance. In the posterior nerve root secondary acute degeneration of fibres occurs, and is followed by sclerosis of varying amount. In the peripheral nerves degeneration appears and then subsides, and is replaced by sclerotic changes.

Degenerated fibres are also seen in the spinal cord, on the inner side of the tip of the posterior horn, and can be traced upwards. The changes in the posterior nerve roots, peripheral nerves and spinal cord are shown well by Marchi's method of staining. When a certain ganglion is diseased "the eruption most frequently lies over a

definite tract of skin, which may be called the normal area from which fibres enter that ganglion" (Head). Hence the distribution of herpes zoster has been employed for determining the areas of skin supplied by the various posterior root ganglia. The area of distribution of the herpes has been mapped out during life, and in a few cases, in which post mortem examination has been obtained, the posterior root which has supplied this area has been determined by the degeneration presented when stained according to Marchi's method (Head and A. W. Campbell).

Head has mapped out a series of segments or areas occupied by the eruption, and the sensory nerve supply of these areas he has attributed to definite posterior nerve roots and ganglia. In ten out of nineteen consecutive areas, Head and Campbell have found by post mortem examination, that the posterior nerve root had been correctly numbered (localised). These areas overlap one another to a very variable extent, "yet in no case does the zone of overlap equal in extent more than one half of the area above and below, whilst in many cases it is considerably less."

In cases of herpes zoster in the areas which are numbered in Head's diagrams as cervical 3, 4, dorsal 2, 4, 6, 7, 8, 11 and lumbar 1, changes have been found in the ganglion of the corresponding posterior nerve root (i.e. in the nerve roots respectively which Head regarded as the sensory nerve supply of these areas).

The Distribution of Sensation with Reference to the Pain of Visceral Disease (Referred Pain).—Many years ago Dr. Ross, of Manchester, pointed out that in visceral disorders there is not only local pain, but also pain referred along the distribution of the somatic nerves which come from the same part of the cord as the sensory sympathetic fibres to the organ affected.

Thus in Fig. 57 at *r c* are the rami communicantes passing from the united anterior and posterior roots to the sympathetic ganglionated cord. From this cord the splanchnic nerves pass to the viscera—(*S p* indicates nerves to the stomach). The splanchnic nerves of the stomach for example are derived from certain dorsal nerves. When the splanchnic nerves of the stomach are irritated, the irritation is conducted to the posterior roots of the dorsal spinal nerves, with which they are connected, and "on reaching the grey matter of the posterior horns it diffuses to the roots of the corresponding somatic nerves (*S o*) and thus causes an associated pain in the territory of distribution of these nerves" somatic pain (Ross).

The whole subject was carefully investigated by Dr. James Mackenzie, of Burnley, and directly afterwards by Head. The results of these and other observations support Ross's theory. In visceral disease pain and tenderness occur in the skin area supplied by afferent root cells of the same spinal segment in which the *visceral* afferent root cells are situated. Referred pains and areas of tenderness in visceral disease

are distributed, not strictly in the course of peripheral nerves, but in the skin areas supplied by the spinal ganglia. In other words, the various viscera receive their sensory fibres from the same segment of the spinal cord as the somatic sensory roots along which the pain is referred in visceral disease. Further, the segmental distribution

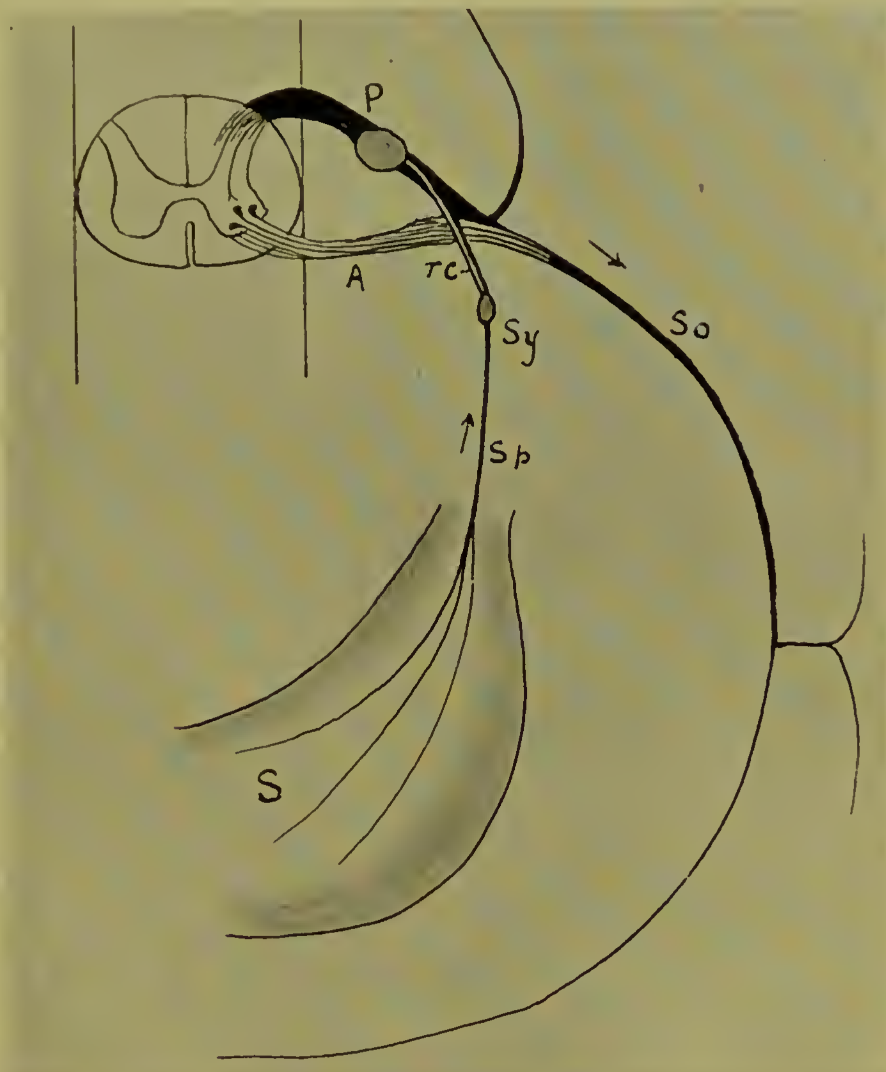

FIG. 57.—Diagram indicating position of referred Pain.
A = Anterior, B = posterior spinal nerve roots.
Sy = Sympathetic ganglionated cord.
Sp = Splanchnic nerve of stomach.
So = Somatic nerve.
S = Stomach.
Impulses passing from the stomach by *S p* to the spinal cord are referred as pain along the somatic branch or peripheral spinal nerve (*S o*).

of the skin fields of the spinal ganglia being known, it is possible by observing the position of the referred pain and tenderness to infer what viscus is diseased. In this way Head has worked out the localisation of the afferent nerve supply to the various viscera, and also the segmental or skin distribution of the various sensory nerve roots.

The following table shows Head's conclusions as to the afferent nerve supply of the various viscera :—

Nerve Root and Spinal Segment.	Heart.	Lung.	Stomach	Intestine.	Rectum.	Liver and Gall Bladder.	Kidney and Ureter.	Bladder, Mucous Membrane and Neck.	Bladder, over distension and ineffectual contraction.	Prostate.	Epididymis.	Testis.	Ovary.	Appendages.	Uterus in contraction.	Uterus, lower segment and internal os.
D 1. .	×	×														
D 2. .	×	×														
D 3. .	×	×														
D 4. .		×														
D 5. .		×														
D 6. .			×			× ?										
D 7. .			×			×										
D 8. .			×			×										
D 9. .			×	×		×										
D 10 .				×		×	×			×		×	×		×	
D 11 .				×			×		×	×	×			×	×	
D 12 .				×			×		×	× ?	×			×	×	
L 1 .							×		×		×			×	×	
L 5. .																
S 1 . .								× ?								? ×
S 2 . .					×			×								×
S 3 . .					×			×								×
S 4 . .					×			×								×

Head's charts show the distribution of the various segmental areas. These are based (1) on the areas of cutaneous tenderness in visceral diseases, (2) on the distribution of the eruption in cases of herpes zoster, (3) on the areas of analgesia in organic diseases of the spinal cord and sensory nerve roots. Head does not regard these areas as absolutely correct, but as only approximately so.

Peripheral sensory nerves.—Head, Rivers and Sherren conclude that the afferent fibres in the *peripheral nerves* can be divided into three systems.

1. Those which subserve *deep sensibility* and conduct impulses produced by pressure and the movements of parts. The fibres of this system run mainly with the motor nerves and are not destroyed by division of all the sensory nerves to the skin.

2. Those which subserve "*protopathic*" sensibility and respond to painful cutaneous stimuli and to extremes of heat and cold; but they do *not* enable us to form any definite appreciation of the locality of the spot stimulated. In any peripheral nerve the distribution of the protopathic fibres usually overlaps greatly the area supplied by fibres of the adjacent nerves. (Similar visceral protopathic fibres pass to the internal organs.)

3. Fibres which subserve "*epicritic sensibility*"—fibres which endow the skin with sensibility to light touch. They conduct the impulses which enable us to localise the position of cutaneous stimuli, to discriminate two points, and to appreciate minor degrees of heat and cold. In the larger peripheral nerves the distribution of these fibres overlaps little.

When the sensory impulse reaches the spinal cord, it "becomes shunted into tracts devoted to the conduction of impulses, grouped in a way different from that found in the peripheral nerves. It is no longer a question of protopathic, epicritic, or deep sensibility, the tracts in the central nervous system are devoted to the conduction of impulses concerned with pain, heat, cold and touch" (Head, Rivers and Sherren).

The Viscero-motor reflex and Viscero-sensory reflex of Dr. James Mackenzie.—James Mackenzie has written many articles on the pain associated with visceral disease and its explanation; he has also given a detailed account of "viscero-motor reflexes," and "viscero-sensory reflexes" and disputes Head's view that the viscera have a "protopathic" sensibility.

[The views expressed, in these recent papers on pain, by J. Mackenzie and Head are of much interest, but further reference to them has not been made on account of limited space and for several other reasons. They concern the general pathology and symptomatology of diseases of the nervous system rather than the special section of it to which this book is devoted, and further the author hesitates to express his own opinions and criticism since they are not based on any extensive observations on the subject, but only on simple experiments which any layman could make.]

Localisation of Reflexes.—The *reflexes* are of much value in spinal localisation. Though a complete transverse lesion may be associated

with loss of all of the reflexes below the lesion, on the other hand, the presence of the various reflexes in spinal diseases indicates that the "reflex arcs" are not destroyed at certain levels of the cord. The following table indicates the segments of the cord at which the reflex arcs of various reflexes have been localised Lesions of the cord at these segments cause loss of the reflexes localised therein.

Cervical	5	Scapulo-humeral reflex.
	6–7	Triceps reflex and wrist jerk.
Dorsal	4–7	Epigastric reflex.
	8–12	Abdominal reflex.
Lumbar	1–3	Cremasteric reflex.
	2–4	—Knee jerk.
	5 – Sacral 1	Tendo Achillis jerk and ankle-clonus.
Sacral	1–2	Plantar reflex.

Relation of Spinal Roots and Segments to Spines of Vertebræ.—The vertebral spines may be counted, if the patient is not too stout : but three landmarks are worth remembering. The 3rd dorsal spine is on a level with "the commencement of the spine of the scapula": and the 7th dorsal spine on a level with the lower angle of the scapula (L. Holden). A line drawn transversely across the lumbar region connecting the highest point of the crest of each iliac bone would cross the spinous process of the 4th lumbar vertebræ (Gumprecht) or the interval between the 3rd and 4th lumbar spines.

The following is the relation of the superficial origins of the spinal nerve roots to the spinous processes of the vertebræ given by Sir V. Horsley (*System of Medicine*, edited by Prof. Clifford Allbutt, vol. vi.)

Superficial Origins of Spinal Nerve Roots.		Apices of Spinous Processes of Vertebræ.
Cervical 1		
2	——→——	1 Cervical
3	....	
		2
4	....	
		3
5	....	
		4
6	——→——	5
7	....	
		6
8	....	

Dorsal	1	———→—	7
	2	....	
			1 Dorsal
	3	....	
	4	———→—	2
	5	———→—	3
	6	....	4
	7	....	
			5
	8	....	
	9	———→—	6
	10	....	7
	11	....	8
	12	....	
			9
Lumbar	1	———→—	10
	2	....	
	3	———→—	11
	4	....	
	5	....	
Sacral	1	....	
			12
	2	....	
	3	....	
	4	....	
	5	....	
Coc		....	
			1 Lumbar.

(*b*) **Spinal Localisation in the Transverse Direction.**—In the transverse section of the spinal cord we have the various tracts of white matter; the grey matter, with its anterior and posterior horns; and the anterior and posterior nerve roots. With respect to transverse localisation of spinal lesions, the following facts of practical importanee may be mentioned:—

In the *crossed* (*or lateral*) *pyramidal* tracts motor fibres pass down from the brain to the anterior horns of grey matter of the eord at various levels as already deseribed. Lesion of these tracts at any point causes paralysis of the *whole* of the muscles below the lesion, which are innervated by the nerve fibres in the traet at the seat of the disease. Secondary degeneration occurs in the erossed (or lateral) pyramidal tracts below the lesion down to the lowest part of the cord. To the clinieal group of symptoms, associated with diseases of the crossed pyramidal tracts, the name of spastic paralysis or spastic paraplegia is given. This is not the name of a disease, but is the term applied to a group of spinal symptoms, occurring in many affections of the spinal cord. These symptoms are (1) loss of power in both legs, with or without affection of the arms; (2) rigidity of the affected limbs on passive movements; (3) increase of the reflexes; increase of the knee-jerk, ankle-clonus, extensor type of the plantar reflex (Babinski's reflex)—the last reflex may be regarded as a sign of affection of the crossed pyramidal tract; (4) spastie gait, if the patient is able to walk—the toes are scraped on the ground, and the legs are rigid, in walking.

The diseases eausing spastic paraplegia are tabulated on p. 261.

Diseases of the *posterior columns* of the cord (columns of Burdach and Goll) are associated with ataxia, with loss of reflexes in some cases, and often with sensory disturbances, chiefly of the muscular sense.

The most important spinal diseases causing ataxia are tabulated on p. 291.

Disease of the *anterior horns of grey matter* causes paralysis of muscles supplied from the affected part of the horn, whilst muscles supplied from the anterior horn below or above the diseased region are not affected. The paralysed muscles are flaccid, and undergo atrophy: the reflexes are lost at the affected level, and the muscles often present the reaction of degeneration on electrical examination, when the lesion is a severe one.

The diseases in which there is *atrophy* of muscles with loss of power are tabulated on p. 196.

Disease of the *posterior horns* of grey matter causes loss of sensation to pain and temperature, with loss of reflexes at the affected part, whilst tactile sensation may be normal. These sensory symptoms are caused most frequently by syringomyelia, spinal hæmorrhage, localised myelitis, and spinal syphilitic changes affecting the posterior grey matter.

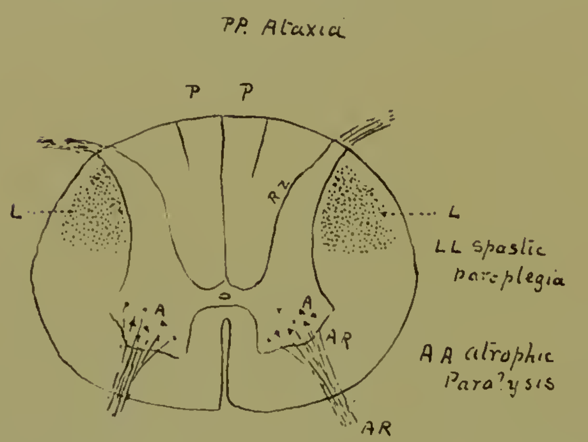

FIG. 58.—Spinal Localisation in the transverse direction.
PP = Posterior columns, lesion causes ataxia.
AA = Anterior horns, lesion causes atrophic paralysis.
LL = Lateral pyramidal tracts, lesion causes spastic paraplegia.

Nerve Roots. Lesion of an *anterior nerve root* causes flaccid paralysis of the muscles which it supplies; atrophy of the paralysed muscles follows. The reaction of degeneration is obtained on electrical examination, and vaso-motor symptoms occur. Lesion of the eighth cervical and first dorsal anterior nerve roots is followed by oculo-pupillary symptoms (*see* p. 86.)

Lesion of a *posterior nerve root* causes anæsthesia in the region of its distribution. Irritation of the root causes pain and hyper-æsthesia. The symptoms caused by lesion of the posterior nerve roots are of great importance in the diagnosis of tumours affecting the spinal cord and its meninges, in caries of the vertebrae, and meningitis.

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Ross, J. *Brain,* vol. x., p. 351.
Mackenzie, James. *Medical Chronicle.* Vol. xvi., p. 293, 1892. *Brain,* vol. xvi., and *British Medical Journal,* June 23 and 30, 1906.
Head, H. *Brain.* Vol. xvi. vol. xvii., and vol. xxviii.

SENSORY NERVES.

Head, Rivers and Sherren, J. *Brain.* Vol. xxviii., 1905.

Localisation of functions in the group of nerve cells of the anterior grey matter.—The nerve cells of the anterior horns of grey matter are arranged in definite groups. In the lumbo-sacral region A. Bruce recognises the following group :

(*a*) Mesial, (*b*) antero-lateral, (*c*) postero-lateral, (*d*) post-postero-lateral, (*e*) central, (*f*) anterior. The position of these groups is indicated in diagram 59. These groups of cells have probably definite functions. It has been already mentioned that when a nerve fibre is divided the cell from which it arises presents certain changes which can be detected by Nissl's method of staining (réaction à distance, *see* p. 41). This method of staining is of great service for demonstrating the functions of various groups of nerve cells.

After removal of the muscles of a limb, as by amputation of a limb or segment thereof, the nerve cells from which their motor nerve fibres arise, show this degenerative change (réaction à distance). At a later period (in man) complete atrophy of the cell may occur, whilst the nerve cells connected with the remaining active muscles of the limb do not show this degenerative change. Thus the functions of various groups of cells can be localised. Fig. 59 indicates functions attributed by Bruce to the various cell groups.

For further details on this subject the reader is referred to the writings of A. Bruce and van Gehuchten, which contains numerous illustrations.

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Gehuchten, van. *Anatomie de système nerveux de l'homme,* Louvain, 1906.

The *localisation* of disease *in relation* to the distribution of the spinal *blood vessels* is described on p. 24.

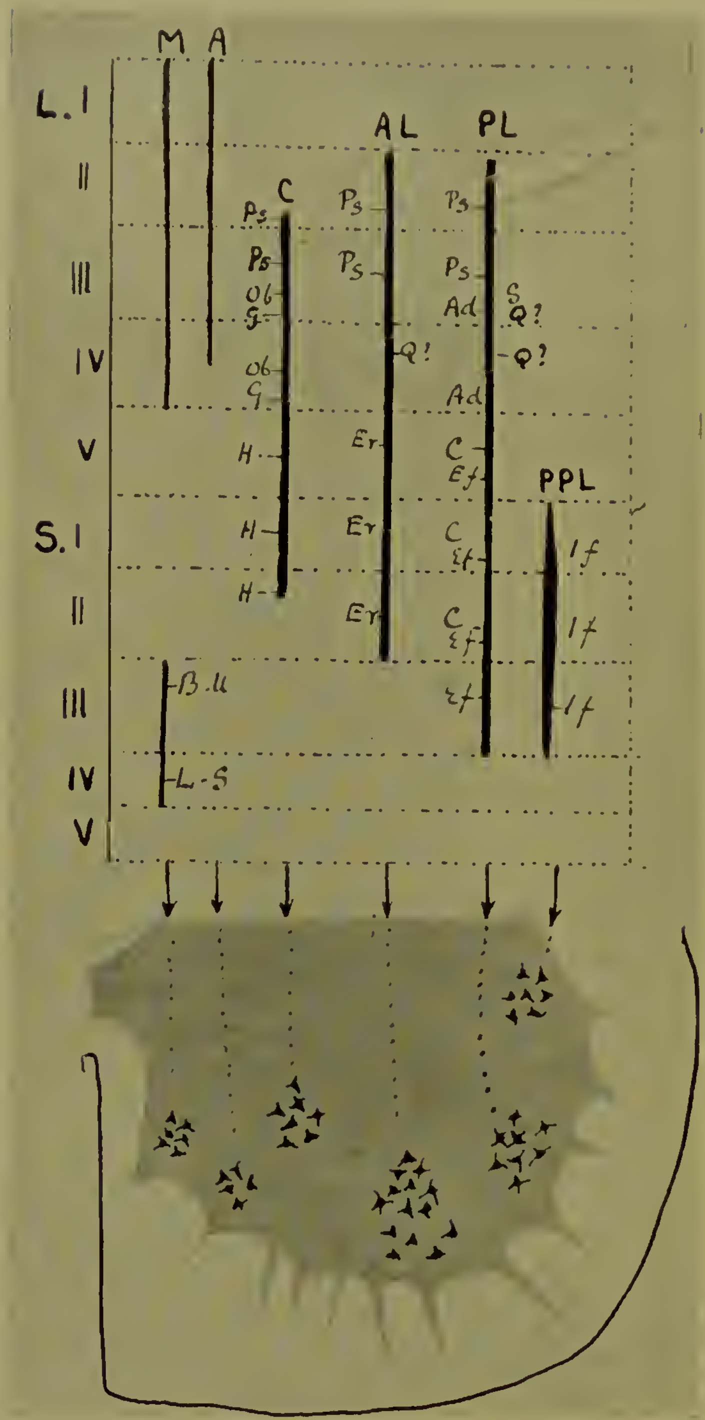

FIG. 59.—Groups of Nerve Cells in Anterior Horn of Lumbo-sacral Region. Motor function indicated (according to A. Bruce). The lower part of the figure may be regarded as the ground plan, the upper part as the elevation. The lower part of the figure indicates the groups of nerve cells in the anterior horn, the upper shows the vertical extent of these cell groups. The figures to the left and the dotted transverse lines indicate the various lumbar and sacral segments.

Cell groups.—M = mesial ; A = anterior ; C = central ; AL = antero-lateral ; PL = postero-lateral ; PPL = post-postero-lateral.

Functions :—BU = bladder and urethra ; LS = levator and sphincter ani : Ps = psoas and iliacus ; Ob = obturator externus ; G = gluteus medius and mininus ; H = hamstrings ; Er = external rotators of thigh ; AD = adductors ; C = calf muscles ; Ef = extrinsic muscles of foot ; S = sartorius ; Q = quadriceps ; If = intrinsic muscles of foot.

Pathological Diagnosis.

Having decided at what region of the spinal cord the disease is situated, an opinion should be formed as to its nature ; i.e. a pathological diagnosis should be made.

Inflammatory and degenerative changes are the most common affections. In some cases, to which the term myelitis is applied clinically, the changes are more of the nature of degeneration (acute parenchymatous degeneration) than inflammation.

Softening of the cord may be the result of inflammation (myelitis), of acute parenchymatous degeneration, or of thrombosis or obstruction in the spinal vessels as a result of syphilis.

In other cases softening is due to compression of the cord by a tumour of the vertebræ, of the spinal meninges, or of the cord itself. Spinal hydatids may produce softening in the same way. But the most common cause of this secondary softening is caries of the vertebræ. In all of these cases very often the first change is œdema of the cord, which is followed by softening or by inflammatory changes.

To all of the forms of softening just mentioned the term myelitis is frequently applied, though a large proportion of the cases are not truly inflammatory in nature.

In chronic degenerative affections the nerve fibres and cells gradually waste, and the neuroglia connective tissue slowly increases. To these chronic changes the name of sclerosis is given.

The *mode of onset* of the symptoms affords some indication as to the pathological nature of the lesion. A very sudden onset (develop-

Onset of Symptoms in Diseases of the Spinal Cord.

Vascular Lesions	*Abrupt.*—(minutes)	Hæmorrhage (spinal and meningeal).	
		Puncture wounds.	
		Compression from fracture-dislocation.	
		Softening of vascular origin, thrombosis ("apoplectiform myelitis").	
	Acute.—(hours or days)	Acute myelitis.	
		Softening from thrombosis	
		Acute anterior poliomyelitis.	
		Acute syphilitic "myelitis."	
Inflammation		Acute meningitis.	
		Landry's paralysis.	
	Sub-acute.—(weeks)	Sub-acute myelitis.	
		Sub-acute anterior poliomyelitis	Compression Myelitis caused by vertebral caries or tumour. Spinal hydatids. Tumours of the meninges and cord. Some forms of spinal syphilis.
	Chronic.—(months)	Pachymeningitis.	
		Chronic meningitis.	
		Some forms of spinal syphilis.	
Degeneration	*Very Chronic.*—(half-years or years)	Amyotrophic lateral sclerosis.	
		Progressive muscular atrophy.	
		Chronic anterior poliomyelitis.	
		Disseminated sclerosis.	
		Tabes dorsalis.	
		Friedreich's disease.	
		Ataxic paraplegia.	
		Combined postero-lateral degeneration of various forms.	
		Syringomyelia.	

ment of symptoms in a few minutes) is characteristic of a vascular lesion—hæmorrhage or vascular obstruction. A very chronic onset, in which many months or years elapse before the symptoms reach their complete development, is characteristic of the chronic degenerations of the cord. Inflammatory affections have usually an acute or sub-acute onset. In compression of the cord (from tumour growths, etc.) the development of symptoms is usually chronic or sub-acute.

The *etiology* and *history* often give useful indications as to the pathological condition. A family history of the affection is often obtained in Friedreich's disease, in the family form of spastic paraplegia, and in the hereditary progressive muscular spinal atrophy of children. In these affections several members of the same family, or several generations may suffer.

As regards the age of the patient, disseminated sclerosis usually commences before the age of forty and very rarely after that age; Friedreich's disease usually begins before the age of twenty; tabes dorsalis commences after the age of twenty, except in the very rare cases of juvenile or hereditary tabes, in which there are generally indications of congenital syphilis, or of syphilitic infection at a very early period of life.

An injury to the back may produce fracture-dislocation and compression myelitis, or it may cause myelitis, spinal or meningeal hæmorrhage, or pachymeningitis.

A history of syphilis is important. A number of forms of spinal disease are due to syphilitic infection (forms of spinal syphilis and tabes dorsalis). Other spinal diseases have no relation to syphilis—as, for example, amyotrophic lateral sclerosis and disseminated sclerosis.

Evidence of tubercular disease at some part of the system is of diagnostic importance, because caries of the vertebræ with compression myelitis, or tubercular tumour of the cord may be secondary to tubercular mischief in another part of the body.

Many cases of myelitis follow acute ailments of an infectious nature—infectious fevers, gonorrhœa, etc.

Severe anæmia is sometimes associated with, and may be the cause of, certain forms of postero-lateral degeneration; or both the anæmia and spinal disease may be due to a primary toxin.

Spinal changes may be the result of chronic poisoning—(ergotism, pellagra, lathyrism, etc.). Disseminated myelitis has occasionally followed carbon monoxide poisoning.

Evidence of tumour growth, or of hydatid cysts, in other parts of the body may give an indication of the nature of a spinal affection.

The localisation of the lesion with respect to the transverse section of the cord is of some service in the pathological diagnosis. If the symptoms indicate an affection chiefly of the lateral pyramidal tracts then the differential diagnosis of diseases causing spastic paraplegia

will require consideration. If the lesion can be localised to the anterior horns of grey matter, the differential diagnosis of the affections causing atrophic paralysis will have to be considered. If the symptoms point to a lesion of the posterior columns, the differential diagnosis of the affections causing ataxia must be made.

If the symptoms indicate a transverse or a unilateral lesion of the cord, the diseases which are known to most frequently cause such lesions will require consideration in the diagnosis.

Unilateral Lesions of the Spinal Cord.

Hemiparaplegia: Brown-Séquard's Paralysis.

Over fifty years ago, by experiments on animals and clinical observations, Brown-Séquard showed that a unilateral lesion of the spinal cord was followed by paralysis on the side of the lesion, and anæsthesia on the opposite side. Also on the side of the lesion the muscular sense was diminished or lost, whilst on the opposite side it was preserved. On the side of the lesion there was hyperæsthesia for pain and slight elevation of temperature. Bladder and rectal symptoms were sometimes present, but were not constant.

Whatever view may be held as to the exact course of sensory fibres, clinical observations in man support, on the whole, the view that *disease* of one-half of the cord causes paralysis on the side of the lesion, and anæsthesia for pain and temperature sensations on the side opposite to the lesion.

To recapitulate, in man a localised unilateral lesion of the spinal cord produces the following symptom :—

A. On the side of the lesion :

(1) Motor paralysis of the leg, with increase of the tendon reflexes. though at first they are diminished. In time the leg becomes spastic. Ankle-clonus and the extensor type of plantar reflex, are often observed.

(2) Loss of the muscular sense (sense of position.)

(3) Slight elevation of skin temperature.

(4) Hyperæsthesia of the skin.

(5) Near the level of the spinal lesion, a narrow transverse band of anæsthesia, in the area of distribution of the sensory nerve root, or roots arising from the diseased portion of the cord. In some cases there is a girdle sensation around one half of the trunk.

(6) Above this anæsthetic band a zone of hyperæsthesia.

B. On the side opposite to the lesion :

(1) Anæsthesia to pain and temperature, and in some cases to tactile impressions.

(2) Motor power, muscular sense, reflexes, and skin temperature normal.

(3) Above the anæsthesia a narrow zone of hyperæsthesia. (*See* Fig. 60.)

When the unilateral lesion is in the cervical region, the arm and leg are

paralysed on the side of the lesion; and there is anæsthesia on the leg and trunk of the opposite side. Oculo-pupillary symptoms may

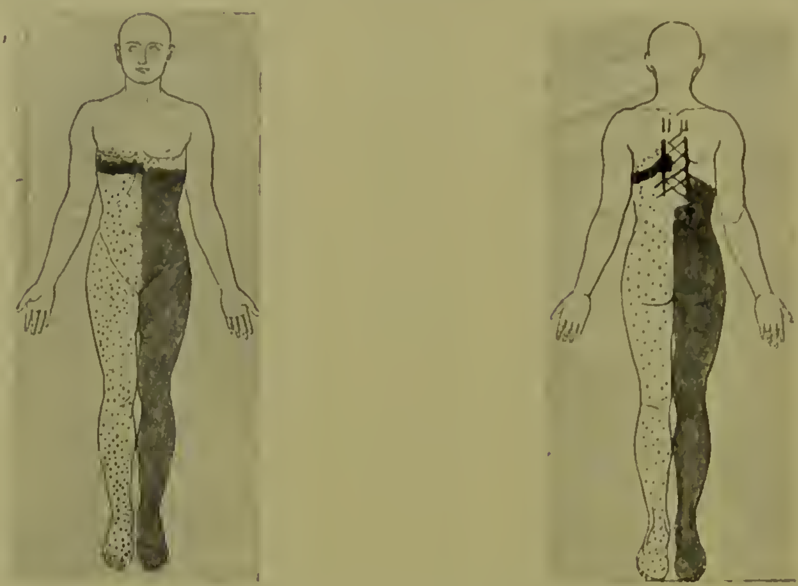

FIG. 60 (A and B).—Unilateral Lesion of the Spinal Coid. The shaded areas = anæsthesia. The dotted areas = hyperæsthesia.

be present on the side of the lesion. When the unilateral lesion is in the lower lumbar or sacral region there is paralysis with sensory dis-

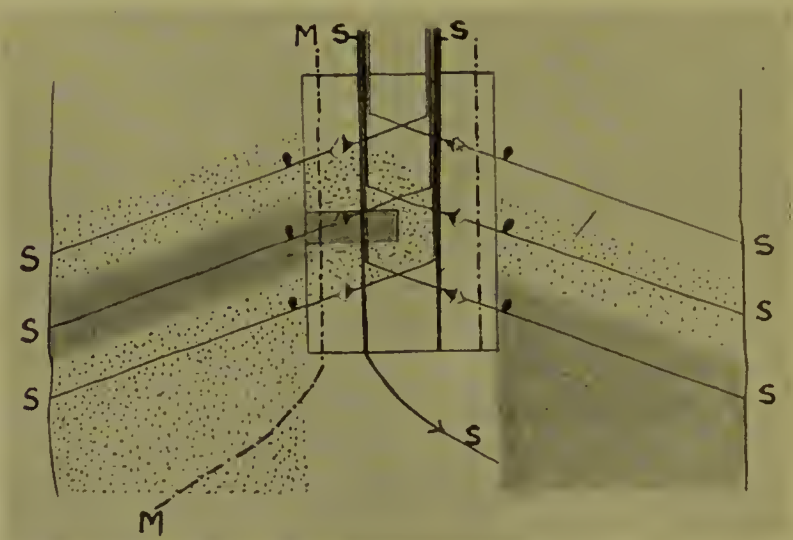

FIG. 61.—Diagram to indicate symptoms in a Unilateral Lesion of the Spinal Cord. Shaded area = anæsthesia; dotted area = hyperæsthesia. M = path for motor fibres and muscular sense; S = peripheral sensory nerves and sensory tracts in cord. The unilateral lesion is indicated in the left half of the cord. The dotted area around the lesion indicates irritation of nerve fibres. The band of anæsthesia on the side of the lesion is caused by destruction of the peripheral nerve S, as it passes into the lesion; the anæsthesia on the opposite side by destruction of the sensory tracts, containing the fibres which have decussated; the hyperæsthesia by irritation of sensory nerve fibres and of the sensory tracts around the lesion (indicated by the dotted area).

turbances in the leg on the side of the lesion, (since the sensory fibres have not decussated or only partially decussated at this level). In

some cases of lumbo-sacral unilateral lesions, recorded by Wernicke and Mann, there has been paralysis and atrophy of the muscles of the leg on the side of the lesion, with anæsthesia of the leg on this side, and, in addition, anæsthesia of half of the scrotum, perinæum, and penis of the *opposite* side.

As regards the explanation of the symptoms in Brown-Séquard's paralysis, it has been already pointed out (p. 52) that probably the fibres conveying the sense of position pass upwards in the posterior columns of the cord without decussating.

Many believe that the tracts for the conduction of sensation of pain and temperature decussate (passing first into the posterior grey matter, and then to the antero-lateral white matter of the opposite side), whilst

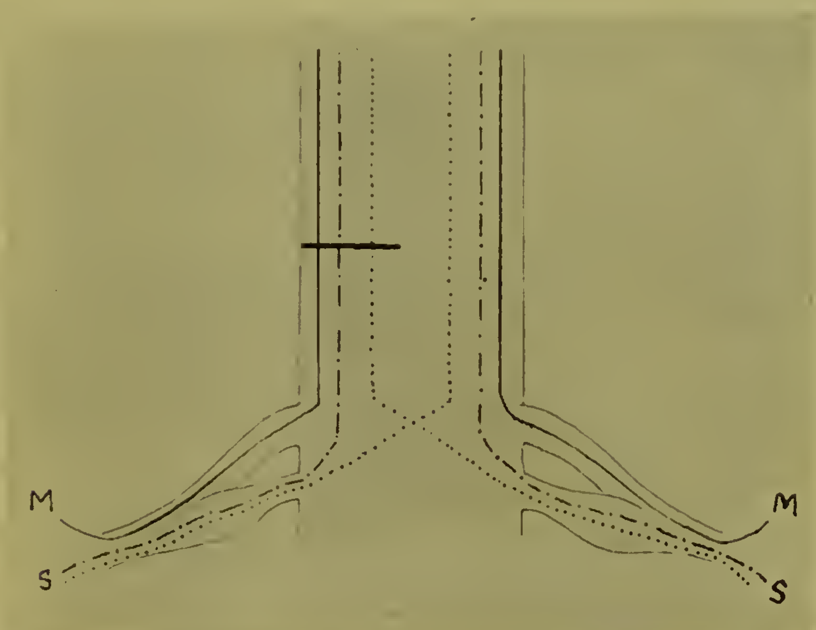

Fig. 62.—Diagram of Spinal Cord to indicate symptoms in a Unilateral Lesion. M = motor fibres of anterior nerve root ; S = sensory fibres of posterior root. Motor fibres = continuous line. Path for impulses of muscular sense and tactile impressions which do not decussate = interrupted line. Path for sensations of pain and temperature and tactile impressions which decussate = dotted line. The unilateral lesion is indicated by a transverse line. This would cause loss of motor power and muscular sense on the side of the lesion ; loss of pain and temperature on tho opposite side (modified after Strümpell).

tactile sensations may be conducted upwards on both sides of the spinal cord (*see* p. 52).

Brown-Séquard's paralysis may be caused by various unilateral lesions—trauma (especially puncture wounds affecting only one-half of the cord), spinal syphilis (meningo-myelitis, gumma and other forms), hæmorrhage, tumour of the cord or its meninges, and very rarely by a patch of sclerosis (disseminated) affecting only one-half of the cord.

In most cases the symptoms of Brown-Séquard's paralysis are only partially developed. Often the anæsthesia is limited to the sensation for pain and temperature, whilst tactile sensation is preserved. Also Brown-Séquard's paralysis is often temporary only, and soon complete

paralysis and anæsthesia develop on both sides. The prognosis is more favourable in cases due to injury, syphilis, or hæmorrhage.

References to most of the important articles will be found in papers by Turner, *Brain*, vol. xiv., and by Warrington, W. B. *Medical Chronicle*, April, 1903.

Transverse Lesion of the Spinal Cord.

Transverse lesion of the spinal cord causes paralysis and anæsthesia below the lesion. In dogs the reflexes are increased and spastic paralysis is produced ; in monkeys the results are variable as regards the reflexes ; in man, after sudden division of the cord, as in the decapitation of criminals, it has been possible to obtain the knee-jerks for 90 seconds.

In disease of the cord in man it was formerly taught that a transverse lesion caused increase of the knee-jerks and spastic paralysis of the legs, when the lesion is above the trophic centre of the leg muscles and these centres are not affected by pathological changes. (The trophic centres for the muscles of the legs are the nerve cells of the anterior horns of the lumbo-sacral cord ; the trophic centres for the muscles of the arms are in the cervical and first dorsal segments.)

When the trophic centres of the muscles are involved in the lesion, then atrophy of the paralysed muscles occurs. Also, when there is an affection of the reflex arc of nerve fibres, on the integrity of which a definite reflex depends, that reflex is no longer obtained.

The bladder and rectum are paralysed in complete transverse lesions of the cord at all regions (*see* p. 85).

The upper limits of the paralysis and anæsthesia indicate the upper extent of the lesion. When reflexes are obtained below the lesion, then the reflex arcs on which they depend, directly or indirectly, have not been damaged by the downward extension of the disease.

In many cases of transverse lesion of the cord the whole of the nerve fibres are not destroyed at the seat of the disease, i.e. the lesion is not completely transverse, and the older descriptions just given would usually hold good. For cases of this kind—incomplete transverse lesions—the following are the symptoms which are usually observed.

When the lesion is at the lower or mid-dorsal region there is paralysis of both legs, paralysis of the bladder and rectum, anæsthesia or impaired sensation in the legs and trunk, the upper limit of which varies with the level of the lesion. Immediately above the anæsthesia there is a narrow zone of hyperæsthesia, and at this level there is often a girdle sensation. Usually the legs become rigid, the knee-jerks increase, ankle-clonus and the Babinski type of plantar reflex appear.

When the lesion is in the upper dorsal region, the intercostals are also paralysed.

When the lesion is in the lower cervical region certain muscles of the arms are paralysed and soon become atrophied. Anæsthesia extends down the inner side of the arms or over a larger area, according to the level of the lesion. The trunk and legs are also anæsthetic. The atrophied

arm muscles present the reaction of degeneration on electrical examination. The intercostals, the legs, and the bladder and rectum are also paralysed as in a lesion of the upper dorsal region, and the paralysis of the legs is spastic.

When the upper cervical region is affected the diaphragm usually becomes paralysed and death occurs from asphyxia.

When the lesion is in the lumbar region the legs are paralysed and flaccid, the knee-jerks are absent, the paralysed muscles atrophy markedly and show the reaction of degeneration, the bladder and rectum are paralysed. Large bedsores are very liable to develop over the sacrum. The upper limit of the anæsthesia varies with the extent of the lesion (*see* diagram 56). In these cases the reflex arcs are broken down in the lumbar region.

FIG. 63. — Anæsthesia of the Legs and Trunk in a Transverse Lesion of the Cord (shaded area). Above the anæsthesia is a zone of hyperæsthesia (dotted area).

But Bastian has pointed out that a total transverse lesion—one destroying every nerve fibre and nerve cell in the transverse lesion—causes *loss* of the reflexes and flaccid paralysis below the lesion, even when the lesion is above the lumbar part of the cord, and the latter region is not involved in the disease.

[A few cases have been recorded, however, in which the knee-jerks have persisted, and the subject is one which has been much debated during the last ten years. W. B. Warrington has discussed the whole question carefully, and has pointed out, that whilst in general the symptoms described by Bastian are caused by complete transverse lesions of the cord, there are exceptions. When the disease is of a slowly progressive nature, the reflex functions of the cord may be retained, and the muscular tone may be unimpaired. Also in some cases very severe symptoms, approximating to those generally found after complete transverse lesions, may be present, and yet the anatomical lesion is slight or incomplete.]

As a general rule, it may be stated that the activity of the deep reflexes usually distinguishes partial from complete division of the cord.

Collier has recently made a careful study of the subject of *total* transverse lesions in man, and from pathological and clinical observations he comes to the following conclusions :—

As the result of *total* transverse lesion of the spinal cord in man, not only are the knee-jerks and other deep reflexes permanently abolished in the region supplied by the portion of the cord below the lesion, but in addition the muscles waste and lose their faradic excitability, and the sphincters lose their tone, the only sign of self action remaining in the isolated part of the spinal cord being the occasional presence of

certain of the skin reflexes in much reduced degree. These phenomena occur in the absence of any recognisable structural change in the anterior horn cells, anterior nerve roots and peripheral nerves of the paralysed region. In these cases, for a certain time after their disappearance, the knee-jerks may be again obtained subsequent to long continued faradisation of the lower limbs. This result may be of service in localising the lesion—the return of the knee-jerk after faradisation indicating that the lumbo-sacral enlargement and its nerves are not involved in the lesion.

After the occurrence of a total transverse lesion, there may be temporary retention of urine, and there may be reflex evacuation for a time ; but the persistence of these phenomena seems to indicate that the lesion is not total. Constant dribbling with a small bladder has been the rule in total transverse lesions.

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Collier. *Brain*, 1904, p. 38

Clinical Groups of Spinal Diseases.

Spinal diseases may be divided into four groups, with regard to the nature of the most prominent symptoms. This division is useful in considering the differential diagnosis, and also enables us to remember better the most important symptoms of the various diseases.

These four clinical groups are :

1. Diseases in which the prominent symptoms are those of a *transverse lesion* of the cord.

2. Diseases in which the prominent symptoms are those of *atrophic paralysis.*

3. Diseases in which the prominent symptoms are those of *spastic paresis* or *paralysis.*

4. Diseases in which the prominent symptoms are those of *ataxia.*

After the consideration of these groups, the various forms of inflammation of the meninges of the cord will be described, and finally the lesions of the cord and its meninges due to injury, tuberculosis, and syphilis.

SECTION VII

DISEASES IN WHICH THE PROMINENT SYMPTOMS ARE THOSE OF A TRANSVERSE LESION OF THE SPINAL CORD

In many diseases of the spinal cord the symptoms are those of a transverse lesion; and from the mode of onset of these symptoms, or from the presence or absence of certain other symptoms, the pathological diagnosis can be made.

The symptoms of a transverse lesion are present in the following diseases: the diagnostic peculiarities as to mode of onset and the additional symptoms are indicated in the table:

Symptoms of Transverse Lesion of the Cord.

Causes.	Onset.	Additional Symptoms.
1. Spinal hæmorrhage, hæmatomyelia	abrupt	
2. Apoplectiform myelitis	abrupt	
3. Acute transverse myelitis	acute	+ Slight premonitory symptoms—numbness, tingling: slight febrile symptoms.
4 Intra-medullary spinal tumour	gradual	
5. Extra-medullary spinal tumour * (meningeal)	gradual	+ Root symptoms.
6. Vertebral tumour	gradual	+ Root symptoms: sometimes bone symptoms.
7. Vertebral caries	gradual	+ Root symptoms and bone symptoms.

* Including spinal hydatid.

Symptoms of a *unilateral* spinal lesion, paralysis on one side and anæsthesia on the other (or Brown-Séquard's paralysis), may be caused by punctured wounds, spinal hæmorrhage, tumours of the cord or its meninges, and by some forms of spinal syphilitic lesions.

Symptoms of localised irritation or destruction of nerve roots ("root symptoms") may be caused by tumours of the spinal meninges, by vertebral caries and tumour, by pachymeningitis, and by some forms of spinal syphilis.

ACUTE MYELITIS AND ACUTE SOFTENING (OR MYELO-MALACIA)

(Derivation, Gr. μυελὸς=marrow; itis=suffix indicating inflammation or disease.)

The term myelitis should be applied only to inflammation of the spinal cord; but many diseases which are now regarded as chronic degenerations, were formerly described as myelitis.

The most common form of spinal disease to which the term myelitis

is now applied is the so-called "compression myelitis," which is caused by compression of the spinal cord owing to caries of the vertebræ, spinal tumour, or other lesions. In such cases the changes in the spinal cord are very frequently not truly inflammatory in nature, but are due to œdema followed by degeneration of nerve elements.

In a second group of cases to which the term myelitis is applied the disease of the cord is produced by syphilis or tuberculosis. But in most of these cases the pathological changes have special characters, often there are gummata or tubercles in the diseased parts, or there is diffuse gummatous or tubercular infiltration with vascular changes. It is better not to apply the term myelitis to these cases, but to describe them as forms of syphilis or tuberculosis of the cord.

In a third group of cases there is no compression, mechanical cause, or primary gross lesion to which the cord disease is secondary. The cord changes are diffuse in nature at the part affected; the nerve elements are diseased and destroyed without implication or exclusion of special groups of cells or tracts of fibres; and the changes are acute or subacute in their onset. To such cases the term primary or idiopathic myelitis is applied. (When the changes are limited to the grey matter of the cord the disease is spoken of as poliomyelitis.)

In some of these cases the pathological change is really acute softening or degeneration owing to the action of a toxic substance on the cord, or owing to obstruction of spinal vessels. But in other cases the vessels are surrounded by cell infiltration; the nerve tissues are infiltrated with cells; and there are the definite appearances of inflammation. These are cases of true myelitis.

Excluding compression myelitis, syphilitic and tubercular cord disease, and poliomyelitis there is then a group of cases to which the term primary myelitis may be conveniently applied in clinical medicine.

The diseases termed myelitis may be therefore classed in the following manner:—

A. Affections often termed "myelitis" which ought to be distinguished from true myelitis:—

I. Compression "myelitis."

II. Syphilitic and tubercular "myelitis"—cord diseases with special characteristics.

B. Affections to which the term myelitis may be applied in the clinical sense:—

III. Primary acute myelitis.

Pathologically this group includes:—

(*a*) True inflammatory conditions—true acute myelitis.

(*b*) Acute degeneration of toxic origin.

(*c*) Acute degeneration following thrombosis or obstruction of spinal arteries.

All of the three conditions *a*, *b*, and *c* may lead to softening of the cord, though this is not always the terminal change.

Etiology.—Often the cause of myelitis is obscure. Many cases follow febrile diseases such as typhoid, typhus, smallpox, malaria and influenza. Occasionally myelitis has followed the septic infection caused by a whitlow. It has occurred in rare instances after gonorrhœa and occasionally has followed parturition.

It is the disseminated form of myelitis which is specially liable to follow infectious diseases.

Syphilis may cause a myelitis resembling that following fevers, as regards the pathological changes. But in many cases of so-called "syphilitic myelitis" the changes are of a special character, or they are due to softening from vascular obstruction.

Tuberculosis is said to produce a myelitis in rare cases, but usually special changes are present.

Muscular over-exertion and strain, the lifting of heavy weights, sexual excess have all appeared to be exciting causes in rare cases.

Some cases have followed exposure to wet and cold; others have followed injury to the spine.

In experiments on animals, myelitis produced by the injection of organisms is caused more readily after injury to the vertebral column or after exposure of the back to cold.

Often no cause of the myelitis can be detected, but febrile symptoms are present at the onset.

Pathological Anatomy and Histology.—The meninges of the cord are often congested. To the naked eye the cord itself may not present any definite changes, but in cases of localised myelitis the diseased area is often softer to the touch of the finger than other parts. In old standing cases the affected area may be firmer.

On section, at the diseased part, the cut surface may appear hyperæmic, and sometimes minute hæmorrhages are seen. In other cases the cut surface is grey, and darker than in the normal condition. Often it is not possible to distinguish between the grey and white matter.

In some cases the cord substance is soft, diffluent, or cream-like on section. The softening may be red, grey, or yellow, according to the amount of blood pigment and altered blood pigment at the softened part. If a small particle of the cord substance be removed from the softened part and examined under the microscope it presents numerous large cells containing fat globules—compound granular cells.

After hardening the cord in Muller's fluid, on section the diseased areas are much paler than the normal parts.

In sections stained according to Weigert's method the diseased parts are paler than the rest of the section; when carmine or aniline blue black are employed as stains, the diseased parts are more deeply stained.

Microscopic examination at the region of the myelitis shows breaking down and destruction of nerve elements indiscriminately, without any special localisation of the changes to groups of nerve cells or tracts of fibres.

When the changes are slight the nerve fibres are swollen; the axis-cylinders are enlarged, and the myelin sheath is narrow and distended. At a more advanced stage both axis-cylinders and myelin sheaths break up into granular masses, which may be finally absorbed; empty spaces are then left in the neuroglia tissue.

The nerve cells at first present the changes of chromatolysis; at a later period they are granular, the nuclei are indistinct, the processes disappear and the cells break up into granular fragments.

The diseased area may be infiltrated with leucocytes and compound granular cells, with here and there a few red blood corpuscles; and mixed with the cell elements are granules produced by the degeneration of nerve cells and fibres. In some cases the cell infiltration of the tissue is the chief change, and nerve cells and fibres are only slightly affected at an early period.

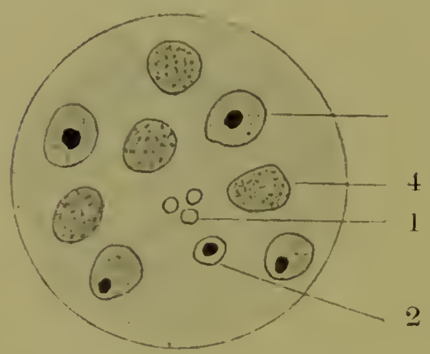

FIG. 64.—Cells from Exudation in Acute Myelitis.
(1) Red blood corpuscles.
(2) Nucleated small cells (leucocytes).
(3) Nucleated large cells.
(4) Compound granular cells.

The neuroglia in time proliferates, and spaces are left in it from which the nerve fibres have disappeared. Sometimes the neuroglia appears amorphous or granular; in old-standing cases it is firmer, its nuclei are increased in number, and numerous "spider cells" or Deiter's cells are present.

Compound granular cells are seen in the spaces of the neuroglia network and in large numbers around the vessels. They are very numerous in many recent cases; but rarer in old-standing cases.

The vessels are often dilated and full of blood. Around the vessels are clusters of nuclei; and the nuclei of the capillaries are increased. The perivascular sheaths are usually greatly distended and full of compound granular cells or leucocytes.

At a late period there is extensive and dense proliferation of neuroglia —sclerosis—at the region of the myelitis. This dense fibrous tissue contains numerous spaces from which nerve fibres have disappeared (*see* Fig. 76).

Above the myelitic area there is ascending degeneration or sclerosis in the posterior median and direct cerebellar tracts and in the tract of Gowers; below the lesion, descending degeneration or sclerosis in the crossed and direct pyramidal tracts (as described on p. 32).

As already mentioned the finer pathological changes vary considerably in cases which are diagnosed clinically as primary acute myelitis.[1]

In the one pathological group of cases—acute *parenchymatous degeneration*—the microscopical examination shows a marked swelling of nerve

[1] In the following description the author is much indebted to the work of Schmaus and Sacki.

fibres, affecting both the axis-cylinders and the medullary sheaths. The latter appear much dilated and cyst-like, and stain faintly with Weigert's stain. The whole section ,of the diseased part presents a pale appearance. The markedly swollen axis-cylinders are sometimes homogeneous and deeply stained, in other cases pale, granular and degenerated. In sections stained according to Marchi's method, the swollen fibres are stained deep black. Longitudinal sections show that the axis-cylinders are often degenerated in short segments, rolled spirally, and surrounded by swollen fragments of degenerated medullary substance. The neuroglia fibres are changed to swollen thick homogeneous or granular strands, which often stain deeply. In advanced cases the altered neuroglia has a peculiar melted appearance ; also the neuroglia cells are often swollen. In most cases there is dilata-

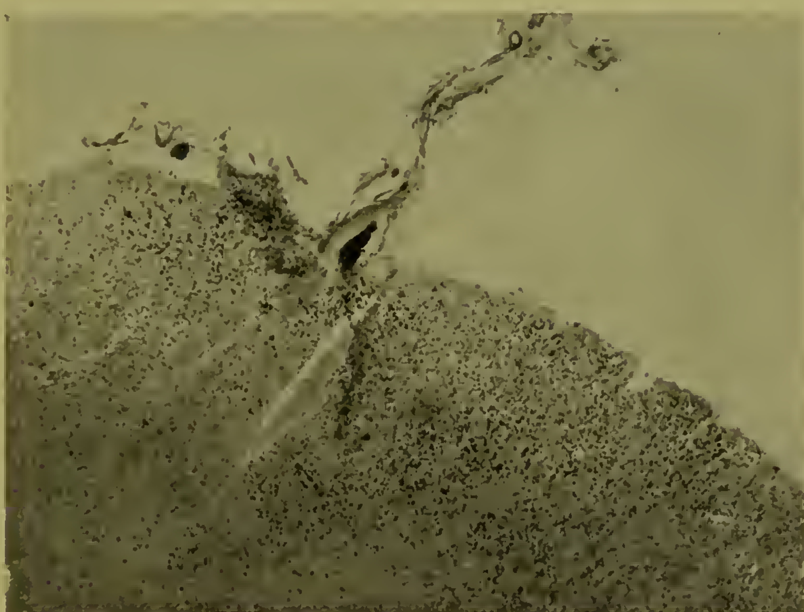

FIG. 65.—Acute Myelitis (Marchi's stain). Degenerated nerve fibres (black).

tion of the small arteries and veins, accumulation of homogeneous or slightly granular transudation masses in the interstitial tissue, exudation of isolated clusters of leucocytes in the walls of the vessels, and free in the tissues, and scattered compound granular cells in the latter. Schmaus and Sacki regard the changes as the result of inflammatory œdema. This form of myelitis is therefore characterised by swelling and œdema of the tissues and degenerative changes in the nerve elements. It may be described as the parenchymatous degenerative form of myelitis —just as we describe a nephritis as parenchymatous, when the changes are chiefly in the parenchyma of the kidney.

In a second group of cases, the *vascular* changes are the most marked. Microscopical examination shows as the chief change a marked small cell infiltration, which mainly occurs in the neighbourhood of the vessels, and forms spots or streaks of cell infiltration in the cord substance, or in rare cases a diffuse infiltration. The cell infiltration is most

marked in the lymphatic sheaths of the blood vessels, but is also seen in the surrounding tissue; often the pericellular spaces of the ganglion cells are filled with round cells. Some of the cells have polymorphous nuclei, others are multi-nuclear leucocytes, and others round cells with a single round nucleus. In addition to the cell infiltration there is hyperæmia and serous infiltration of the tissue, with swelling of the structures, as in the parenchymatous form. The degeneration may be more, or less, extensive than the region of cell infiltration. The degeneration is often apparently only secondary to the cell infiltration ; not infrequently small hæmorrhages are present. In cases of some duration,

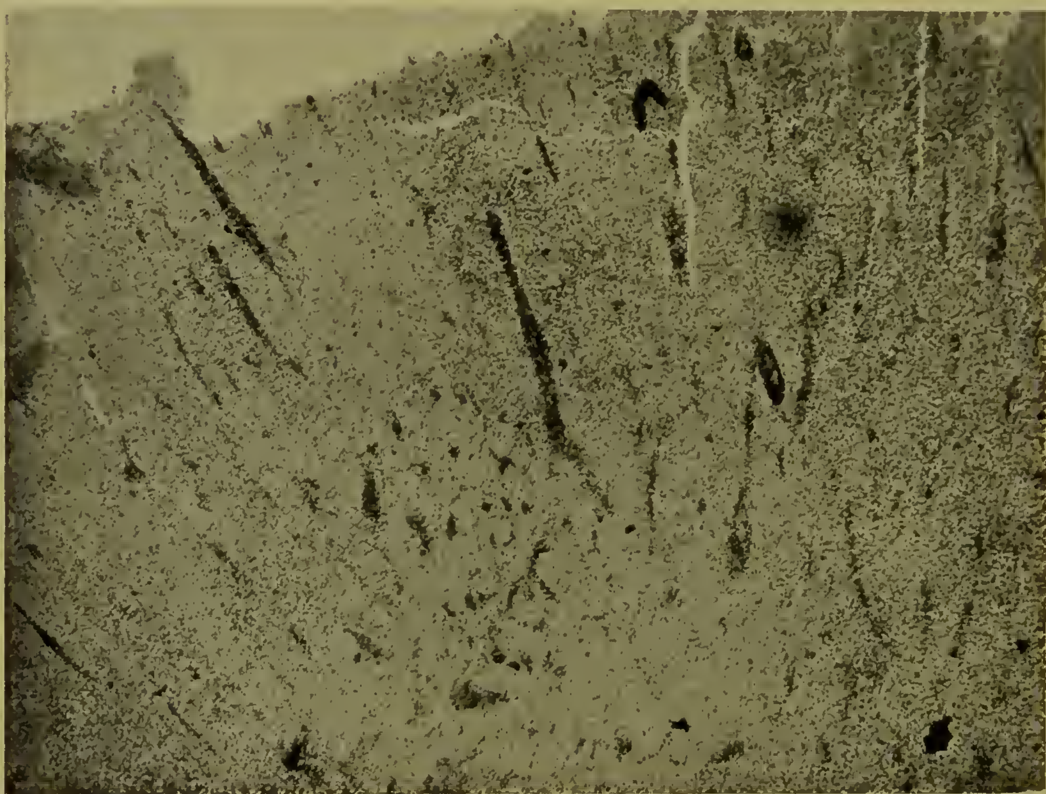

FIG. 66.—Acute Myelitis (logwood stain). Note cell infiltration (black dots). Cells are especially numerous around blood vessels—in the perivascular lymph spaces.

compound granular cells are found along with the round cells in the cell infiltration. The former cells contain fat and myelin drops, and blood pigment. The lymph sheaths of the vessels may in time be filled with compound granular cells.

In a third group of cases the changes are those of typical *softening* and the tissues are infiltrated with compound granular cells. In the softened parts, in addition to the compound granular cells, there are fragments of axis-cylinders, granules of medullary substance, myelin drops, and granular detritus. The vessels present cell infiltration of their walls and especially distension of the lymph sheath with granular cells, fat and detritus. These three forms of myelitis are not sharply separated and often transitional forms occur. Softening may follow

the two forms of myelitis just described, or it may follow thrombosis of a spinal artery.

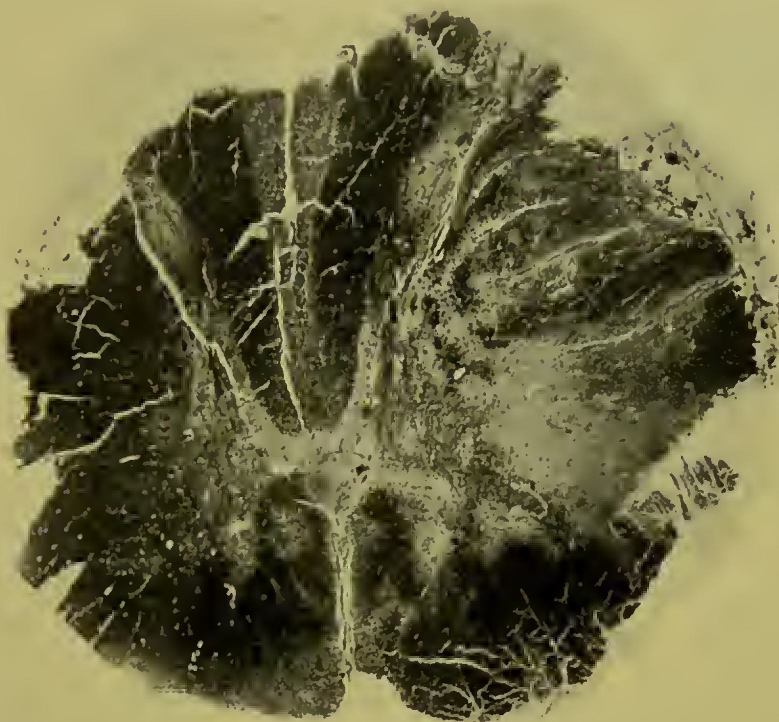

Fig. 67.—Disseminated Myelitis, cervical region (Weigert's stain). Patches of Myelitis pale.

Forms of Myelitis.—The myelitis may extend across the cord at one level forming a *transverse* myelitis. Above and below the lesion

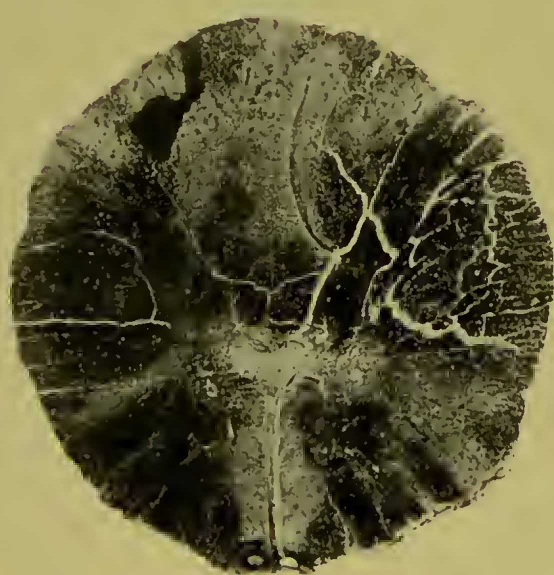

Fig. 68.—Disseminated Myelitis (Weigert's stain). Dorsal region: patches of Myelitis pale.

there is usually no myelitis. Sometimes there are small scattered patches of myelitis immediately above and below the transverse lesion, but

none elsewhere. This transverse form of myelitis is most common in the dorsal region.

In another form—*disseminated myelitis*—there are irregular scattered

Fig. 69.—Disseminated Myelitis, Dorsal region (Weigert's stain). Small patches of Myelitis pale.

patches of myelitis, the margins of which are usually not sharply defined. In disseminated myelitis there is sometimes optic neuritis which is due to pathological changes in the optic nerves exactly resembling those

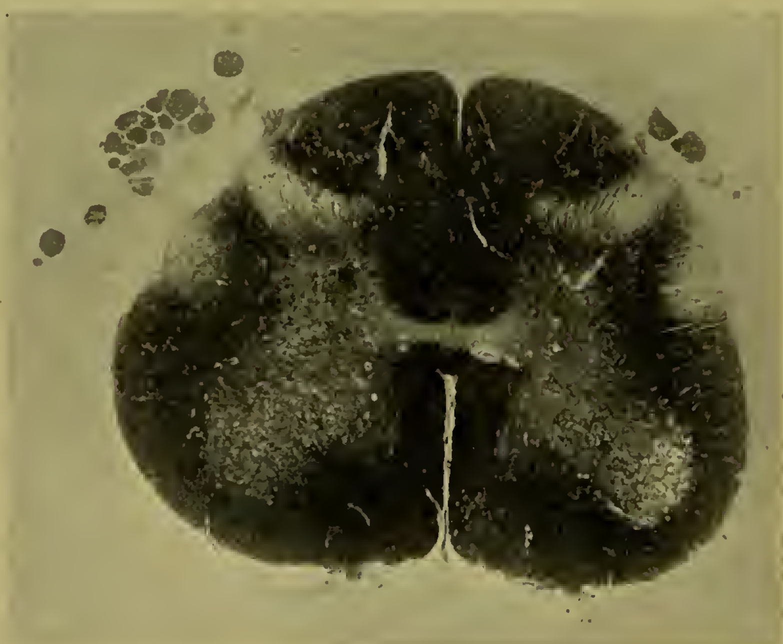

Fig. 70.—Lumbar Region of Cord (Weigert's stain). Descending degeneration in the crossed pyramidal tracts, below extensive disseminated myelitis in the dorsal region. Degenerated tracts pale.

in the spinal cord in nature. Occasionally inflammatory patches are found in the medulla, pons, and brain as well as in the cord.

The patches of inflammation are scattered about in a most irregular manner. These patches have a well marked vascular character; the small blood vessels in the diseased areas are dilated; their walls and their adventitial and perivascular lymphatic sheaths are crowded with round cells; for some distance around the vessels is an area infiltrated with round cells and compound granular cells; very often a greatly dilated blood vessel is situated in the centre of the patch, and the myelitis often extends in streaks along the course of the vessels. The patches of myelitis at the periphery of the cord are often wedge shaped (with the apex directed inwards) and correspond to the distribution of the small peripheral arteries.

The vessels often show marked cell infiltration of their walls, or in the adventitial lymph sheath, before entering into the cord substance. This may be well seen in the anterior median fissure.

FIG. 71. — Transverse Section of Optic Nerve from the case of disseminated myelitis figured in Figs. 67–70. During life there was optic neuritis. The nerve shows irregular inflammatory patches (paler parts in the section).

The nerve fibres and cells at the affected area are degenerated. In cases examined a considerable time after the onset of the disease the nerve elements are entirely broken down in the diseased patches, the tissues are infiltrated with compound granular cells, the cord substance is softened, and minute hæmorrhages are occasionally present. At a later period the diseased patches become sclerosed and the neuroglia connective tissue is increased. But there are several points of difference between the pathological changes in disseminated myelitis and disseminated sclerosis. In disseminated myelitis the margins of the patches are not sharply defined, whilst in disseminated sclerosis in many patches,

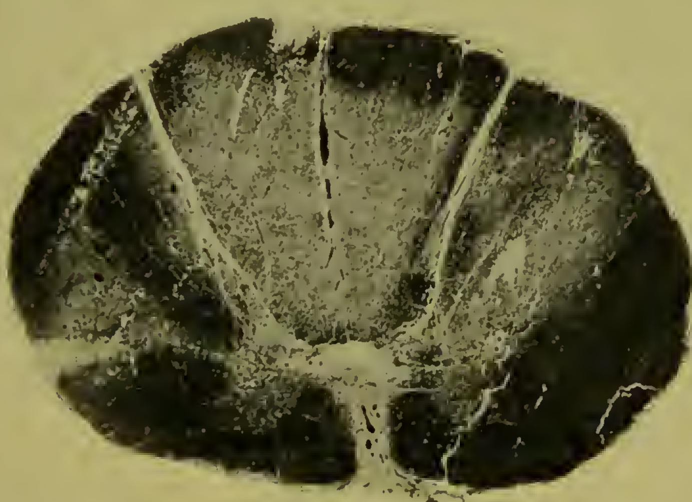

FIG. 72.—Disseminated Myelitis Weigert's stain (second case). Cervical region. The patches of myelitis are pale.

the diseased area is very sharply limited. The disseminated myelitis often follows the course of the vessels in streaks. In disseminated myelitis, in the diseased area, the axis-cylinders are degenerated as well as the myelin sheath, and the nerve cells are also degenerated; whilst in patches of disseminated sclerosis often many axis-cylinders and nerve cells persists, at least for a long period. In disseminated myelitis there is ascending degeneration above the lesion, and descending degeneration below it; whilst in many cases (probably in most) of disseminated sclerosis these secondary degenerations do not occur.

Other forms of myelitis are the acute *central* myelitis, in which the

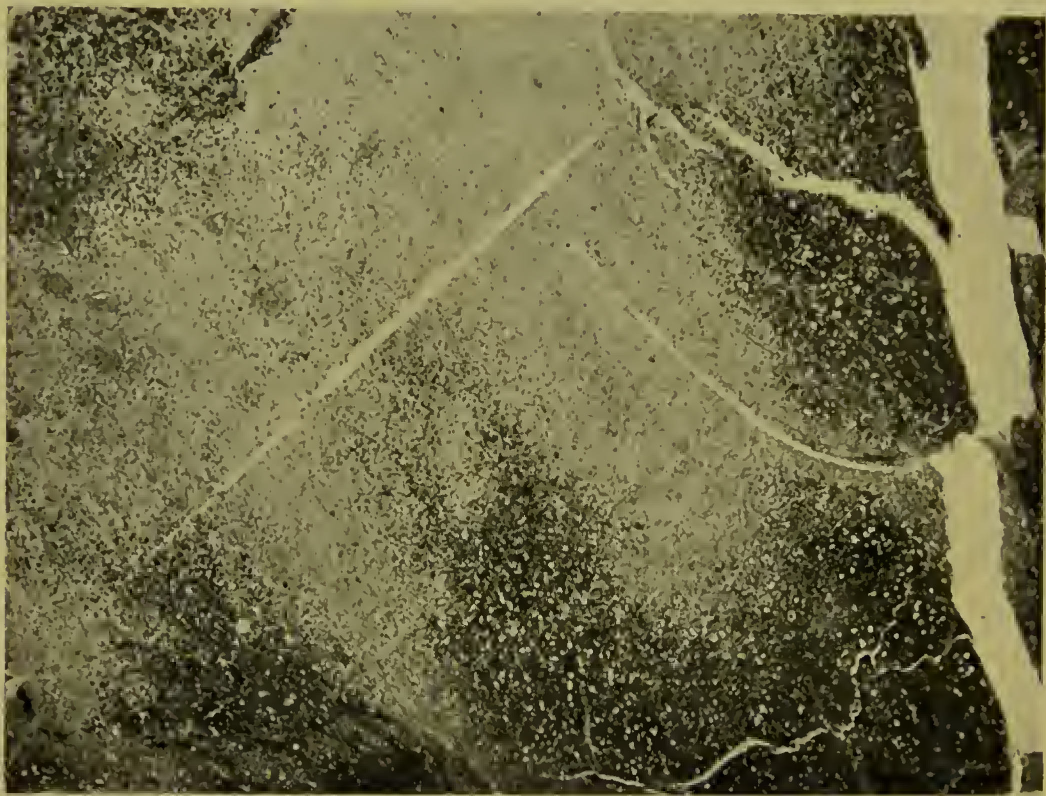

FIG. 73.—Disseminated Myelitis (Weigert's stain). High power of Microscope. The pale areas are patches of myelitis. These show an absence of nerve fibres, which in the normal parts are stained black.

changes affect chiefly the central grey matter of the cord, and *diffuse* myelitis which extends from one primary focus over the greater part of the cord.

Meningo-myelitis is a rare form in which the meninges (pia and arachnoid) and the surface of the cord are both inflamed. The condition may follow injury, septic infection from wounds; it has occurred in infectious diseases and fevers, pneumonia and malignant endocarditis; and the condition is occasionally met with in the epidemic cerebro-spinal meningitis.

Gonorrhœal infection has been followed in rare instances by spinal meningitis or meningo-myelitis; more rarely by disseminated myelitis. In several cases examined bacteriologically no gonococci have been found

in the cord, and the connexion with gonorrhœa is probably indirect. In the inflamed urethra other organisms develop besides the gonococci, and the toxic products formed by these may be the cause of the cord changes (i.e. the condition may be due to secondary infection).

Myelitis produced Experimentally.—Numerous experiments have been made on animals (especially in France) in order to determine whether myelitis can be produced by the injection of micro-organisms into the system. In most of the experiments acute degeneratons have been produced rather than myelitis; but Marinesco and others have produced true myelitis.

Myelitis has been produced experimentally in animals by injection of the pus micrococci, the pneumococcus, the micro - organism of erysipelas, the bacillus coli communis, the bacillus pyocyaneus, and several other organisms. The lesions produced have presented the appearance of simple degeneration, or there has been small-celled infiltration, or infiltration with compound granular cells at the seat of the lesion; also transitional forms have been produced.

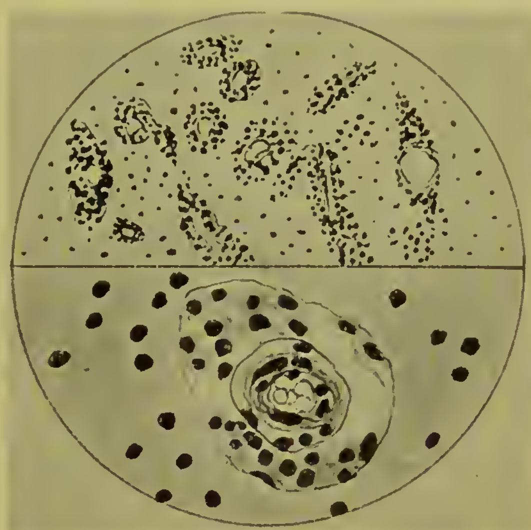

Fig. 74.—Blood Vessels in Disseminated Myelitis. The upper half of the circle shows blood vessels surrounded by cell infiltration (low power). A small vessel is seen in the lower half (more highly magnified): the adventitial sheath is dilated and contains a few cells; the perivascular sheath is dilated and contains many cells.

According to Marinesco cultures of many micro-organisms are capable of producing myelitis when injected into animals, but the more powerful are streptococci, pneumococci and the "toxin" or organism of hydrophobia. Marinesco has produced myelitis by the injection of micro-organisms (1) into a blood vessel distant from the spinal cord; (2) into a blood vessel which directly supplied the spinal cord; (3) into a nerve—such as the sciatic nerve; (4) into the arachnoid cavity. The first method produced myelitis very rarely; the second produced poliomyelitis; the third meningo-myelitis; the fourth meningo-myelitis at the seat of injection. By exposure of the spinal column to the action of cold, and by injury to the back the experimental lesions were intensified.

Myelitis has been produced by the injection of bacterial *toxins* free from organisms. It appears, as if the toxins in some cases act on the vessel wall, in some cases directly on the nerve parenchyma, in others on both.

Marinesco has shown that micro-organisms may disappear from the cord after a few days, and usually they cannot be detected in the cord even when myelitis has been produced experimentally by the

injection of cultures of bacteria. It is not surprising, therefore, that micro-organisms have so often been absent from the cord in man, since the autopsy is not usually obtained until some weeks or months after the onset of the disease. But the following have been detected in the foci of myelitis: streptococci; staphylococci; pneumococci; and a special diplococcus. Marinesco examined six cases of acute myelitis for micro-organisms. In two he found streptococci, in a third pneumococci, in a fourth a bacillus similar to that of anthrax; in a fifth no organism could be found (but the examination was not made until three months after the onset of the acute myelitis). Micro-organisms were not found in a case of poliomyelitis.

Strümpell has recorded the results of examination of the fluid obtained by lumbar puncture in two cases of myelitis. In one the myelitis followed a whitlow; and the fluid obtained by lumbar puncture was turbid and contained numerous staphylococci. Strümpell thinks that probably this organism was the exciting cause of the myelitis. In the second case the lumbar puncture fluid was clear and sterile.

It is probable that when myelitis is due to the action of micro-organisms it is very frequently the result of a mixed infection. Many believe that often the myelitis is not due to the direct action of bacteria but to their toxins.

We may conclude, then, that there is much pathological and experimental evidence in favour of myelitis being caused in many cases by micro-organisms or toxins produced by micro-organisms. The fact that myelitis so often follows infectious ailments is also in favour of this view. In the disseminated myelitis the irregular and wide distribution of the lesions, and the affection of the optic nerves in some cases, is suggestive of a blood change.

The marked perivascular changes in many cases of myelitis is suggestive of the irritation of the endothelial cells of the small vessels and perivascular lymphatics by some toxic substance in the circulation.

* * * * *

It is probable that many cases described as myelitis are really due to spinal softening produced by obstruction of blood vessels. Cases are on record in which pathological examination has revealed the presence of spinal softening due to obstruction of small arteries by thrombi, with or without changes in the walls of the vessels.

The writer has shown that so-called syphilitic myelitis may be due to softening caused by thrombosis in small spinal blood vessels, the walls of which present syphilitic disease (*see* p. 392).

As Sir Wm. Gowers points out, it is conceivable that thrombosis may occur in a minute spinal blood vessel, and that the initial lesion may ultimately disappear in the intense inflammation which it excites.

In some cases of very sudden onset it appears probable that the initial change has been thrombosis in minute spinal vessels, though post-mortem such thrombosis may not be detected.

Recently several writers have suggested that all cases of so-called "myelitis" are really due to softening and not to inflammatory changes. But certainly this view is not justified by pathological examinations.

In many cases the vascular and perivascular changes are very well marked. The adventitial and perivascular sheaths are filled with round cells and there is cell infiltration in the tissues around the vessels. The blood vessels are dilated and filled with blood. The changes in the nerve elements are most marked just around the vessels; and they are widely distributed; also the vessels may be surrounded with round cells before they enter the cord substance. The changes in the nerve element may be slight in comparison with the cell infiltration. No thrombosis can be detected. All these changes point to a primary inflammatory condition.

In softening from thrombosis there is a greater tendency for all the tissues at one area to be degenerated together, i.e. for softening or degeneration to affect indiscriminately all the various histological structures.

It is in the disseminated myelitis that the inflammatory nature of the changes is most decided, and these form a considerable proportion of the cases of so-called "primary myelitis."

At the present time we may conclude, therefore, that pathological examination furnishes clear evidence that the *affection diagnosed clinically as acute myelitis is in some cases inflammatory, in other cases due to acute degeneration or softening from toxic conditions or from arterial thrombosis.*

The true inflammation, true myelitis, is characterised pathologically by marked dilatation of the blood vessels, by extensive infiltration of the adventitial and perivascular lymph sheaths and the surrounding tissues with round cells; whilst in spinal softening these changes are absent or slight.

Probably most of the cases described as disseminated myelitis are truly inflammatory; whilst many of the cases described as transverse myelitis, in which the spinal changes are limited to one level of the cord, are really due to softening. It remains to be decided by further pathological study whether the former cases are always inflammatory in nature and the latter generally due to softening.

Symptoms of Transverse Myelitis—The most common form of acute myelitis is *transverse* myelitis. The symptoms are those of a transverse lesion of the cord of acute onset.

Prodromal symptoms, chiefly numbness and tingling in the legs, are generally present for a day or two, or for a few days.

General symptoms, such as headache, slight pyrexia, slight shivering, and loss of appetite occur at the onset. Sometimes there is a slight girdle sensation of short duration. Dull pain in the back may be present, but it soon disappears, and is not a prominent symptom. Vertebral pain is almost always absent.

The legs become weak and heavy; the loss of power rapidly increases; and in a few hours the legs become paralysed. The paralysis reaches its height in a day, or in a few days. The bladder often becomes paralysed along with the legs, in some cases before the affection of the legs.

In a rare group of cases the onset is more rapid—*apoplectiform* myelitis; the paralysis reaches its height in one or two hours as in spinal hæmorrhage. These cases are difficult to distinguish from spinal hæmorrhage; but the more sudden the onset and the fewer the prodromal symptoms the greater the probability of spinal hæmorrhage; the more distinct the prodromal symptoms the greater the probability of myelitis. In rare instances the post-mortem examination has revealed myelitis, and yet the symptoms have developed as suddenly as in hæmorrhage. Possibly such cases have commenced in thrombosis of a spinal vessel.

Occasionally the onset of myelitis is *subacute*, and several days or a few weeks elapse before the paralysis reaches its height.

Chronic myelitis is extremely rare. Most cases regarded as chronic myelitis are really due to some other form of spinal disease. But very rare cases of myelitis are on record in which spastic paraplegia has developed gradually, and afterwards anæsthesia and bladder symptoms have appeared.

I. *Dorsal Myelitis.*—This is the most common seat of the disease, and the symptoms, when fully developed, are those of a transverse dorsal lesion.

The following description applies to the more common form of dorsal myelitis in which the transverse lesion is probably not complete.

Both legs are paralysed partially or totally. At first the reflexes may be lost; but if the dorsal region only is affected and the lumbar region free, they usually soon return; both the superficial reflexes and the knee-jerks become increased in a short time, and ankle-clonus develops on each side. The plantar reflex is usually of the extensor or Babinski's type. At an early period the paralysed muscles are flaccid (when the reflexes are lost), but in course of time the legs usually become rigid, and a spastic paraplegia develops. When the lumbar region is not implicated there is usually no definite atrophy of the leg muscles, though slight wasting from disuse may occur.

If the changes should extend downwards into the lumbar region, then the knee-jerks disappear and the leg muscles atrophy.

The sensory symptoms vary according to the severity of the lesion. Sometimes there is complete anæsthesia; but more frequently there is impairment of sensation to tactile and painful impressions and to the sense of temperature in the legs and trunk. The upper level of the anæsthesia varies with the level of the dorsal region diseased: it may reach to the umbilicus, to the epigastrium, or to the nipples as described in the account of transverse lesions of the cord.

Sometimes there is loss of sensation to tactile impressions only: or to pain and temperature only. Occasionally sensation is normal or

very little affected. At the upper level of the area of anæsthesia there is often a narrow zone of hyperæsthesia ; and the application of a hot sponge over this zone produces a sensation of pain.

The bladder and rectum are usually affected, and sometimes bladder symptoms occur before the paralysis. At first there is frequently retention of urine. In other cases the bladder empties itself periodically in a reflex manner, the patient having no control over its action. Later there is often incontinence, in the form of dribbling from an over-distended paralysed bladder, or constant incontinence without over-distention of the bladder.

At first there may be obstinate constipation, but this is often succeeded by loss of control over the rectum and incontinence of fæces.

Loss of sexual power is common. Sometimes priapism is a troublesome symptom, and the use of the catheter may produce an erection of the penis. Frequently seminal discharges occur without any erection.

The urine soon becomes alkaline ; on standing a deposit of phosphates and ammonium urate often occurs, and bacteria are present in large numbers. The urea frequently decomposes, ammonium carbonate is formed, and the urine has an ammoniacal smell and reaction, even when it is recently passed. Cystitis is very liable to develop, especially when the bladder is paralysed and is allowed to remain over-distended for a long period, or if a catheter which is not quite aseptic should be introduced into the bladder. When cystitis occurs, in addition to the changes in the urine just mentioned, pus is present along with much mucus.

At the early period the temperature of the paralysed legs is often one or two degrees above the temperature of other parts of the body : but at a later stage the surface temperature of the paralysed legs is often a degree lower than the body temperature.

Bed sores may form over the sacrum, or trochanters ; in rare cases over the heels. Usually they are superficial, but occasionally they extend deeply, down to the bone. Bed sores occur much less frequently in dorsal myelitis than in lumbar myelitis. The skin of the limbs is sometimes dry, sometimes covered with sweat.

Occasionally there is slight œdema of the legs, and occasionally effusion into the knee-joints.

In dorsal myelitis the electrical reactions of the paralysed muscles are usually normal, or there is simply diminished excitability (for exception *see* p. 114).

In course of time the paralysed legs usually become very rigid ; in cases of long duration there is often contraction of the adductors of the thighs, so that the knees are pressed together ; and the legs are flexed at the hips and knees and drawn up to the abdomen, the heels often coming almost in contact with the nates. Frequently there are jerky twitchings of the legs, the limbs being drawn up to the abdomen more strongly. These twitchings are caused by any stimulation of

skin of the legs, such as a slight touch, or even by exposure of the legs to cold air or to a draught.

The condition known as spinal epilepsy is met with in rare cases —clonic spasms or convulsive movements of the legs are produced by slight stimulation, such as striking the patellar tendon. These movements can be arrested by flexing the big toe firmly.

When improvement occurs, the sensory symptoms subside more quickly than the motor.

In less severe forms of transverse dorsal myelitis there is simply paresis with diminution of sensation, instead of complete paralysis and anæsthesia.

In the majority of cases of dorsal myelitis, when the lumbar region is unaffected, the reflexes are increased and the paraplegia is spastic as described ; but occasionally the reflexes are lost and the legs are flaccid, though the lesion does not extend to the lumbar region. According to Bastian the latter condition is met with when the lesion is completely transverse ; the former when the transverse lesion is not complete (*see* p. 114).

II. *Cervical Myelitis.*—In this region a transverse myelitis causes (1) all the symptoms of a dorsal myelitis which have been just described—paralysis of the legs (usually spastic) with paralysis of the bladder and rectum. (2) In addition, the intercostals are paralysed. (3) The arms are paralysed, and the extent of the paralysis of the arm muscles will vary with the extent of the lesion in the cervical cord. If the lower cervical region is affected, the paralysed arm muscles are markedly atrophied, and may present the reaction of degeneration on electrical examination (whilst the reaction of the leg muscles is normal or the excitability simply diminished as in dorsal myelitis). The anæsthesia in cervical myelitis affects the legs and trunk and extends to upper part of the thorax and down the inner side of the arms, or over the whole arm, according to the level of the lesion. There is often a little pain in the neck, with rigidity of the neck muscles and retraction of the head.

Pupillary changes (unilateral or bilateral) may be present. They consist of contraction of the pupil with diminution of the palpebral fissure (paralytic symptoms), or dilatation of the pupil, and increase of the palpebral fissure (irritative symptoms). The face is sometimes pale ; in other cases flushed and covered with perspiration. These symptoms are caused by affection of the cilio-spinal centre or of the fibres passing from it to the sympathetic. The pulse is often rapid. Hyperpyrexia occasionally occurs, the temperature rising to 107–110° F. Priapism is an occasional symptom.

Breathing is carried on by the diaphragm only, when the intercostals are paralysed. If the lesion extends upwards the diaphragm is ultimately paralysed, and death occurs from asphyxia.

In myelitis at the highest cervical region there is atrophic paralysis

of the muscles of the neck; the arms are paralysed, but atrophy of the arm muscles may be absent, and both arms and legs may be spastic. Ultimately the diaphragm becomes paralysed and death occurs from asphyxia.

In rare cases of cervical myelitis optic neuritis has been noticed (*see* p. 87).

III. In *Lumbar* myelitis the legs are paralysed, but are not spastic. The paralysed muscles undergo marked atrophy, and the feet are "dropped." Electrical examination reveals the reaction of degeneration in the paralysed muscles. The superficial reflexes of the leg are diminished or absent. The knee-jerks and the tendo Achillis reflexes are lost, and ankle-clonus is not present. (In very rare instances, however, in which the myelitis has affected the upper lumbar region only, the knee-jerk has been absent but ankle-clonus present.)

The bladder and rectum are paralysed; there is incontinence of urine and fæces from the first; but there is dribbling of urine without any period of retention. Bed sores over the sacrum are very liable to occur; they often appear at an early stage of the disease, and are usually extensive and deep. There is anæsthesia of the legs, complete or partial, and the upper limit of the anæsthesia will vary with the level of the lesion (as indicated on p. 98). If the lesion does not extend above the level of the second lumbar segment the anæsthesia will not extend higher than the groin: if the lesion affects the first lumbar segment then there will be anæsthesia on the lower part of the abdomen for a very short distance above the groin (*see* Fig. 56).

When the lesion is limited to the lowest lumbar region the knee-jerks may be present, and the anæsthesia and paralysis limited to the parts supplied from the lower segments of the lumbar or sacral regions.

If only the conus terminalis is affected there is paralysis of the bladder and rectum, with impotence; anæsthesia is limited and can be detected only in the region of the anus, perineum, scrotum, penis, and inner side of the thigh at the upper part (*see* Fig. 56); paralysis is limited to the muscles or a portion of the muscles supplied by the sacral plexus (*see* p. 185).

Other Forms of Myelitis.—There are several rarer varieties which require brief notice.

Disseminated Myelitis.—In this form patches of acute myelitis are scattered about in a most irregular manner in various parts of the spinal cord. In some cases localised patches of acute inflammation are also found in the pons and medulla, or in the optic chiasma and optic nerves.

In most cases there is paralysis of the legs or of the legs and arms. Often the paralytic symptoms are irregular in development. Thus one leg becomes paralysed, then the other leg, and later the arms; or one leg becomes paralysed, then the arm on the same side, and a short time afterwards the other leg and arm are affected. The paralysed legs may be atrophic or spastic, according to the seat of the lesions. If the myelitis

is in the lumbar region the knee-jerks are lost. A girdle sensation is sometimes present if the changes are chiefly in the dorsal region. The bladder and rectum are paralysed as in transverse myelitis. Often the symptoms very closely resemble those of transverse myelitis, and differ simply in some irregularity in the distribution of the paralysis or anæsthesia. A primary myelitis which continues to extend after the first two or three days is probably disseminated. Optic neuritis, which is extremely rare in diseases of the spinal cord, has been frequently met with in disseminated myelitis, and this sign appears to be a point in favour of the disseminated form of myelitis. Occasionally the optic neuritis has preceded the spinal symptoms. As already mentioned, the optic neuritis may be due to patches of inflammation in the optic nerve similar to those in the cord.

Leyden has especially drawn attention to a clinical group of cases of disseminated myelitis, which he has described as *acute ataxia*. In these cases disseminated patches of acute inflammation are found in the cord, in the medulla, pons and cerebral peduncles. The chief symptom is ataxia in all the limbs, of acute onset. The arms are most affected. The movements of the limbs are often slow, and there is frequently slight weakness distributed irregularly, but real paralysis is rare. Sometimes there is tremor of the limbs on movement like that of disseminated sclerosis; also there may be tremor of the head. The pupils are normal. Involuntary movements of the tongue may be present. Nystagmus is sometimes observed, and the speech is often slow, monotonous, and scanning like that of disseminated sclerosis. Usually the bladder and rectum are unaffected. Pain and sensory affections are usually absent or very slight. The skin reflexes are normal or diminished, the tendon reflexes may be increased. Optic neuritis has been observed. The symptoms may subside in a few weeks; or some symptoms may subside whilst others persist. Leyden thinks these cases may finally develop into disseminated sclerosis. Death sometimes occurs from intercurrent diseases. The affection resembles disseminated sclerosis in many points; but it differs in the acute onset and in ataxia being the prominent symptom.

Acute diffuse central myelitis is a form in which there is rapid loss of power and sensation in the limbs. The arms and legs are affected at the same time, or the legs first and the arms soon afterwards, or the arms first and the legs later. The reflexes are lost, trophic changes occur rapidly; the temperature is elevated, and death occurs in the course of a few days.

Hæmorrhagic myelitis is a variety in which well marked hæmorrhage occurs in association with myelitis. There are slight symptoms of myelitis first, and then severe symptoms, caused by the hæmorrhage, develop very suddenly.

Course and Prognosis in Acute Myelitis.—A few cases recover completely after some months or a year or two. Other cases improve in

course of time, but some of the symptoms remain permanently. Often the condition remains stationary for a long period, and death occurs from some complication. The causes of death in acute myelitis are : (1) Bladder complications and their consequences—cystitis, pyelitis, pyelo-nephritis, or multiple abscesses in the kidneys, with secondary pyæmia ; (2) deep bed sores followed by septic absorption and pyæmia ; (3) paralysis of the intercostal muscles, followed by lung affections—œdema, congestion, bronchitis, broncho-pneumonia ; (4) paralysis of both intercostals and diaphragm, and death from asphyxia ; (5) intercurrent diseases such as lobar pneumonia, broncho-pneumonia and bronchitis.

Some cases terminate fatally at an early period of the disease. In cervical myelitis, death may occur in a few days, the spinal changes ascending and causing respiratory paralysis. In dorso-lumbar myelitis, the cord lesion may ascend and death may occur in a few weeks or months.

Prognosis.—The prospects of recovery are greater if the myelitis has followed some infectious disease. Many cases of syphilitic myelitis recover, but other cases occurring during the early secondary stage sometimes run a rapidly fatal course (*see* p. 391).

The prognosis is worse in disseminated myelitis than in transverse myelitis ; it is also worse in cervical and lumbar myelitis than when the disease is limited to the dorsal region. Marked anæsthesia, paralysis of the bladder, the early development of bed sores, the onset of cystitis, paralysis of respiratory muscles, atrophy of the paralysed limbs and persistent loss of the knee-jerks are unfavourable signs.

Diagnosis.—It is most important to diagnose primary myelitis from "compression myelitis," due to vertebral caries, vertebral tumour and meningeal tumour. Mistakes are frequently made ; very often compression myelitis from caries is diagnosed as primary myelitis because the back is not examined. In compression myelitis, from the causes just mentioned, "root pains" at the level of the disease usually precede the myelitis for some time. It is important to examine the back for a prominent vertebral spine, and other signs of caries, or for signs of a tumour growth (*see* p. 157). It is also important to search for evidences of past syphilis and for signs of tubercular disease, and to examine the various organs for indications of tumour growth.

Examination with the X rays is occasionally of service in the differential diagnosis of acute myelitis from caries or tumour (*see* p. 151).

A history of previous infectious disease, or of exposure to cold, initial febrile symptoms, and an acute onset of the affection are all points in favour of primary acute myelitis.

From *spinal hæmorrhage*, acute myelitis is usually diagnosed by the less sudden onset in the latter disease. In spinal hæmorrhage, the paralysis develops in a few minutes ; premonitory symptoms are absent, and there is sometimes pain in the back at first (*see* p. 188). In myelitis

numbness and tingling in the legs or some premonitory symptoms usually precede the onset of the paralysis. The paralysis requires a few hours or days for its complete development. The more sudden the onset the greater the probability of hæmorrhage; the longer the duration of premonitory symptoms the greater the probability of myelitis. There are rare cases, however, of *apoplectiform myelitis* in which the onset is almost as sudden as in hæmorrhage; possibly these cases have their origin in thrombosis of small spinal vessels. In such cases the diagnosis during life is rarely possible.

In *meningeal hæmorrhage* severe pain in the back and symptoms of irritation of nerve roots precede the paralysis.

Myelitis which ascends the cord may simulate *Landry's paralysis.* But in the former sensory symptoms are present; bed sores frequently occur; and the reaction of degeneration is often met with. In Landry's paralysis, sensation is not affected, there are no bed sores, no electrical changes in the paralysed muscles, and the reflexes are lost.

Acute anterior poliomyelitis of the infant and adult causes motor symptoms only; the bladder is not affected; bed sores do not develop; the reflexes are not increased; ankle-clonus and the Babinski type of plantar reflex are absent. In the infant, often one limb only is paralysed.

In cases of dorsal myelitis in which the knee-jerks are present, the diagnosis from *peripheral neuritis* is easy. In the dorsal myelitis ankle-clonus, a spastic condition of the legs, and bladder symptoms are often present: whilst in peripheral neuritis the knee-jerks and the symptoms just mentioned are absent. A greater difficulty occurs in the diagnosis of cases of myelitis in which the knee-jerks are absent (cases of lumbar myelitis, or of myelitis causing a *complete* transverse lesion of the cord). In these cases of myelitis bladder symptoms are present, bed sores very frequently occur, the anæsthesia has a definite upper level (corresponding to the upper level of a definite spinal segment); whilst in peripheral neuritis, bladder symptoms and bed sores are absent, and the upper border of the anæsthesia is not sharply defined. Also in most forms of peripheral neuritis there is more pain in the limbs, with much tenderness of the muscles, and the diaphragm is paralysed before the intercostals (the opposite occurs in myelitis); moreover there is usually a history of alcoholic excess or of one of the other causes of neuritis.

In *hysterical paraplegia*, the possibility of myelitis may require consideration. In hysterical paraplegia the knee-jerks are never lost; there is no incontinence of urine or fæces, (though retention of urine may occur in very rare cases); the plantar reflexes are often absent, and when present are *not* of the Babinski extensor type; also there is no reaction of degeneration in the affected muscles. From myelitis of the lumbar region, the diagnosis is easy, since in this affection the knee-jerks are nearly always lost, there is incontinence of urine and fæces, cystitis and bed sores are very liable to develop, and

the paralysed muscles present the reaction of degeneration. Dorsal myelitis is more liable to be mistaken for hysterical paraplegia. If the myelitic lesion be complete, as already mentioned, the reflexes may be all absent ; but in most cases the lesion is not complete and the knee-jerks are present and increased. Such cases are usually easily separated from hysteria by the spastic condition of the legs, by the presence of true and sustained ankle-clonus, by the presence of the Babinski extensor type of plantar reflex. Also incontinence of urine and fæces and cystitis are common. In both dorsal and lumbar myelitis, the upper limit of the anæsthesia usually corresponds to that of a definite sensory root distribution.

The diagnosis between *acute myelitis* and *acute softening* is scarcely possible clinically (except perhaps, in syphilitic cases). But Langdon thinks that the absence of all premonitory symptoms, the more sudden onset of the paralysis, the absence of fever and chills, and the absence of any history of preceding acute illness, injury, or disability, are points in favour of acute softening (myelo-malacia). Premonitory symptoms, pyrexia, history of acute illness, or of injury would be in favour of acute myelitis.

Treatment.—Some cases of myelitis recover. By more careful treatment, recovery would be more frequently obtained, since very often death is not due to the disease itself, but to some complication.

Complete rest in bed is of course essential. It is best that the patient should rest on the side, and that the back should not be the lowest part of the body. A long back rest or support is of service in maintaining the lateral position. If the disease be extending, the prone position is advisable for a time. Frequent slight changes in the position are desirable to prevent undue pressure on the skin over bony points.

The two complications which most frequently cause death are bed sores and cystitis, with septic infections therefrom. Hence it is most important to do everything possible to prevent these complications occurring. In order to prevent the formation of bed sores, it is best to employ a water bed from the first. The most careful cleanliness is essential, since the fæces are often passed involuntarily. The bed sheets should be changed at once when soiled, and they should be dusted with some non-irritating dusting powder, such as oxide of zinc and starch powder. If the skin should become red, and if signs of a commencing bed sore should appear, the sacral region or suspected part should be washed with spirit lotion, or with eau de Cologne, or with a lotion consisting of 10 grains of tannin in one ounce of rectified spirits, or with a solution of alum. If a bed sore has actually formed, it should be dressed with zinc ointment, carbolized vaseline, or iodoform ointment. In chronic large bed sores, iodide of starch paste is of service. (For composition see Martindale and Westcott's Extra Pharmacopæia.) If the bed sore should become septic, it may be necessary to wash it with an antiseptic lotion containing perchloride of mercury or other disinfectant.

In some cases of very extensive bed sores a permanent lukewarm water bath has been of service.

It is an important practical point in all cases of spinal disease, never to omit to examine the condition of the bladder by palpation of the abdomen. In myelitis, the patient may state that he is passing water all right; but the urine passed may be simply dribbling from an over-distended and paralysed bladder, i.e. the patient may be suffering from retention of urine with dribbling. In such a case, by palpation of the abdomen, above the pubes, the distended bladder would be felt. If the bladder be allowed to remain over-distended, cystitis is very likely to develop, and this may be followed by pyelitis, pyelonephritis, and fatal septic infection.

If there should be retention of urine, or retention of urine with dribbling, the catheter should be used two or three times a day. But the catheter should be kept perfectly aseptic. There can be no doubt that it is extremely difficult to prevent cystitis when the bladder is paralysed, however careful the attention to strict cleanliness of the catheter may be; but on the other hand, it is quite certain, that through a dirty catheter cystitis is often excited or increased.

A soft india-rubber catheter is best for general use. Before being passed, it should be disinfected well by boiling in water, or by carbolic lotion, or some other antiseptic. The orifice of the urethra should be sponged over with a mild antiseptic solution or with Jeyes' fluid, before the catheter is used. These precautions are important whenever a catheter is passed, but are particularly so when the bladder is paralysed. After being used, the catheter should be carefully washed and placed in antiseptic solution till used again. In order to prevent the lodging of any septic matter, just at the tip of the catheter, beyond the eye, it is best to use a catheter which is solid at this part (i.e., solid at the tip just beyond the eye).

One catheter should not be used too long; when it becomes damaged in any way it should be at once replaced by a new one.

When cystitis has actually developed, the bladder should be washed out twice a day with boracic acid lotion (15 gr. to the ounce of water) made luke-warm just before use by the addition of a little boiling water; or a solution of sodium salicylate (30 gr. to the ounce of water) may be used in the same way. When cystitis occurs, urotropin should be given, gr. 7 to 10, three or four times a day in water; or salol gr. 5 or more, three times a day.

When there is constant dribbling of urine, a porcelain bed urinal may be employed for male patients; but by constant contact it sometimes causes irritation and œdema of the end of the penis, and occasionally sloughing occurs. In such cases the genital organs may be surrounded by some absorbent antiseptic wool which is changed frequently.

If the myelitis has followed exposure to cold or an infectious disease, it is advisable at the onset to employ a diaphoretic treatment. The action of the skin should be promoted by surrounding the patient with

woollen blankets; hot drinks should be given; and Oppenheim recommends the conduction of steam by a suitable apparatus under the bedclothes.

Salicylate of soda has been given frequently at the onset, but its value has not been clearly demonstrated; it may be of service in cases following acute or infectious diseases. It is well to give a mild purgative, such as calomel, at first, and to follow it by a mild saline purgative. Afterwards, if the bowels remain constipated, enemata should be employed. Ergot and belladonna were formerly frequently given, but their value has not been proved.

If there should be a syphilitic history, or evidence of a syphilitic origin of the myelitis, anti-syphilitic treatment is indicated—potassium iodide by mouth and mercurial inunctions, or mercury may be given in the form of grey powder in a pill (*see* treatment of spinal syphilis, p. 399). If the myelitis has followed malaria, quinine or arsenic should be given.

When the symptoms have been present for a long period, then general tonics such as strychnine, iron, quinine, and arsenic are often employed. But strychnine should not be given if there is rigidity of the legs with increased reflexes.

For the relief of the painful spontaneous contractions and great rigidity of the legs, which are often such troublesome symptoms at the later stages of the disease, warmth, warm baths, and passive movements of the leg are of service. The warm baths should be of 5 to 15 minutes' duration, and whilst the patient is in the bath passive or active movement of the legs (or both) should be made. Hyoscine hydrobromate has been employed also for the relief of these symptoms, and sometimes with benefit, commencing with a very small dose, and increasing up to $\frac{1}{100}$ of a grain or more, in chloroform water, once or twice a day (by mouth). Starr recommends bromides. Occasionally division of tendons may be necessary.

Electricity should not be applied to the legs if they are spastic. In lumbar myelitis, when the legs are much atrophied and the muscles flaccid, galvanism of the muscles may be of value. Strümpell recommends the constant current applied to the spine (stabile method for 3 to 5 minutes at the region of the disease). Massage is also of service in these cases.

If bronchitis should occur as a complication, ammonium carbonate is of service.

When the patient is improving, passive movements should be employed, and as the legs have a tendency to become flexed at the hips and knees, the passive movements should be in the direction of extension at these joints. An endeavour should also be made to prevent the contractions as much as possible by the position in bed.

When the patient is able to stand with assistance, he should be encouraged to practise walking with the "go-cart" or suitable apparatus.

For paralysis of the bladder, galvanism may be employed, one electrode

being placed above the pubes, the other on the perineum ; also strychnine or nux vomica may be given when the legs are not spastic.

Formerly, local treatment to the back at the region of the myelitis was frequently employed—such as ice bags, hot applications, and counter irritants, painting with iodine, etc. The value of this treatment is uncertain. Counter irritation should certainly not be employed when the skin is anæsthetic, and it should not be applied in regions which are liable to be subjected to much pressure in bed.

We have no specific treatment for myelitis, except in the syphilitic cases ; but, nevertheless, careful treatment is of the utmost importance. In many cases, if complications can be prevented, the cord changes will, in course of time, subside, and recovery, partial or complete, may occur. But only too often, cystitis, with its consequent pyelitis and pyelonephritis, or septic infection from a bed sore, or some other complication, terminates life in a case which would otherwise have recovered partially or completely, had such complication been prevented.

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Abscess of the Spinal Cord.

Spinal abscess is very rare. In some of the cases on record the abscess has followed injury to the back ; in other cases it has been associated with purulent meningitis ; occasionally it has been secondary to suppurative disease or septic processes in other parts of the body —bronchiectasis, gonorrhœa, prostatitis. In cases recorded by Turner and Collier, the abscess was associated with " compression myelitis " or myelo-malacia caused by vertebral caries or tumour. In most cases there has been purulent spinal or cerebro-spinal meningitis.

The exciting causes of the abscess have been staphylocci, streptococci, the diplococcus pneumoniæ, and a form of actinomycosis.

The abscess has been found in the central region of the cord, or in the posterior horns. The longitudinal extent has varied, often it has been great. The abscess has sometimes had a distinct wall.

The *symptoms* are indefinite. Meningitic symptoms precede the

spinal symptoms for some hours or days. The spinal symptoms are :—paraplegia of acute onset, anæsthesia, and paralysis of the bladder and rectum. The symptoms have often resembled those of acute ascending myelitis. Fever or subnormal temperature and rigors have been observed in some cases.

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COMPRESSION MYELITIS

"Compression myelitis" is the name given to the changes in the spinal cord which are produced by several diseases that cause a localised diminution of the space in the spinal canal.

In some cases there is actual compression of the cord itself by the primary pathological lesion ; but this is not always the condition.

The primary pathological lesion is usually of small vertical extent, and the secondary cord changes affect the transverse area of the cord like a transverse myelitis.

The most common causes of a "compression myelitis" are :—

1. Caries of vertebræ (tubercular, very rarely syphilitic).
2. Tumours of the vertebræ.
3. Meningeal and extra-meningeal tumours.
4. Spinal hydatid cysts.
5. Aneurisms eroding the vertebræ and compressing the cord.
6. Chronic hypertrophic pachymeningitis.
7. Fractures and dislocations of the vertebræ.

COMPRESSION MYELITIS FROM VERTEBRAL CARIES.

Etiology.—Vertebral caries may occur at any age, though it is more common in childhood. It is almost always due to tubercular disease. (There are, however, rare forms of non-tubercular caries following injury or due to syphilis.) Very often, but not always, there is evidence of tubercular disease in some other part of the body, such as tubercular disease of the lungs or lymphatic glands, caries of other bones, and occasionally acute miliary tuberculosis.

Sometimes there is a history of injury to the back, which is the exciting cause.

Pathological Anatomy.—The starting point of the caries is usually in the body of the vertebra, more rarely in the vertebral joints and intervertebral cartilages. The disease very seldom begins in the vertebral arch or its spinous process.

Usually only one vertebra is diseased, but sometimes two or more adjacent vertebræ are affected. Any part of the vertebral column

may be affected, but the disease is more frequent in the dorsal than in the cervical or lumbar regions.

There is usually a tubercular osteo-myelitis or periostitis. The

FIG. 75.—On the left is a transverse section of Normal Nerve Fibre (Weigert's stain) Myelin sheath deep black; axis-cylinder pale. To the right are two nerve fibres in transverse section, showing swollen axis-cylinders and dilated (narrow) myelin sheaths (compression myelitis).

vertebræ are infiltrated with tubercular granulation tissue, which leads to breaking down of the bone and formation of caseous purulent material. The vertebral column sinks forwards and often, but not invariably, one or more spinous processes become prominent posteriorly.

The spinal cord at the seat of the disease is sometimes narrowed by compression: but often there is no direct compression of the cord

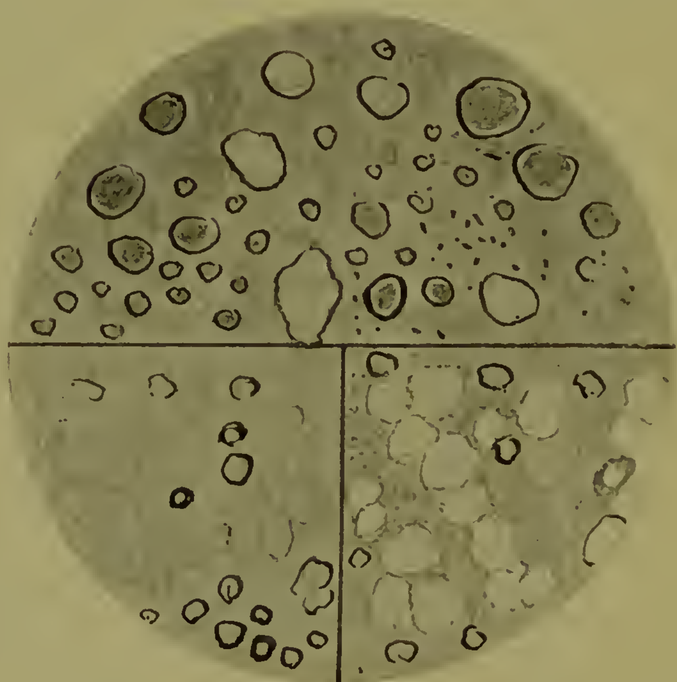

FIG. 76.—Compression Myelitis. Section of cord (Weigert's stain). In the upper half of the circle the section shows nerve fibres with swollen axis-cylinders and dilated medullary sheaths (black). In the lower half of the circle, to the right are the spaces in the neuroglia from which the nerve fibres have disappeared (by complete degeneration); to the left is a portion in which the neuroglia has proliferated, after degeneration of the nerve fibres, sclerosis being produced.

itself. It may be softer than normal at the region of myelitis, but this is not always the case.

Microscopical examination shows, when the changes are slight, that the nerve fibres have swollen axis-cylinders and dilated medullary

sheaths. The latter may stain very faintly with Weigert's hæmatoxylin. The neuroglia may be homogeneous or granular. In more advanced cases nerve fibres and cells are broken down and degenerated, and dilated spaces are left in the neuroglia, which are empty or which contain hyaline or granular masses, fatty material, and compound granular cells. The pericellular lymph spaces and the adventitial lymph sheaths of the blood vessels are dilated; the latter contain numerous compound granular cells. In old-standing cases the perivascular lymph sheaths are also dilated, and may contain round cells. In the neighbourhood of the blood vessels compound granular cells may be found.

The changes in the cord are usually much more marked in the white matter than in the grey. As Schultze has pointed out, they may follow the course of the lateral and posterior arteries, whilst the central arteries remain free. In old-standing cases sclerosis follows the localised cord changes, and secondary, ascending and descending degenerations occur.

The spinal nerve roots are compressed in the vertebral canal or the intervertebral foramina.

As many observers have pointed out, very often the changes in compression myelitis are not inflammatory in nature, and hence the name "myelitis" is scarcely a suitable term.

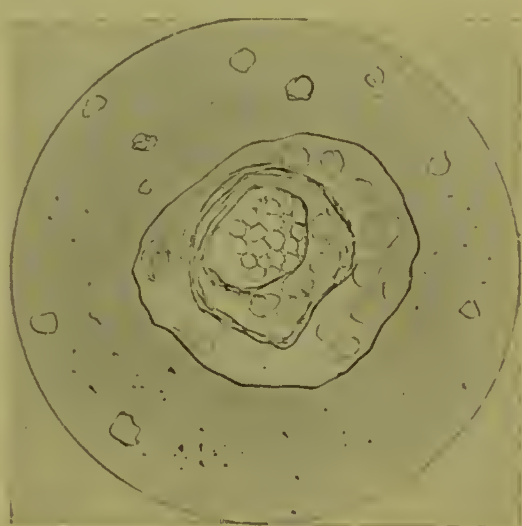

Fig. 77.—Blood Vessel in Compression Myelitis. The adventitial and perivascular sheaths are dilated and contain round cells.

The observations of Schmaus and others have thrown much light on the true pathology of compression myelitis. Occasionally the cord is compressed by *bone*, owing to a displaced vertebra, at the seat of the disease; but this is rare, and is usually due to some injury to the carious spine. In other cases an *abscess* forms at the seat of the carious bone; this is bounded by the vertebral periosteum and the longitudinal posterior ligament, but it may compress the spinal cord. In most cases the *tubercular* disease of the bone *spreads* to the connective tissue and fat outside the dura mater, in the vertebral canal. An external pachymeningitis often develops. This tubercular formation may compress the cord directly, or it may compress the blood vessels and lymphatics and so cause a localised œdema of the cord. Tubercles sometimes spread to the inner side of the dura mater; and the pia mater, arachnoid, and dura mater may all become adherent; in rare cases tubercles invade the spinal cord, and tubercular inflammation may spread along the septa and vessels to the cord.

Œdema from obstruction to the flow of venous blood or lymph is the chief cause of the cord changes, according to Schmaus and Sacki. For a long period localised œdema of the cord may be the cause of the

Fig. 78—Caries of Spine (diagrammatic). On the left is a longitudinal section of the spinal cord and dura mater. To the right is a transverse section of the vertebral column, showing section of spinal cord and dura mater in vertebral canal. The carious part of the vertebra is darker in colour. Note the thickening of the dura mater—external tubercular pachymeningitis, which is causing compression myelitis.

paraplegia—hence the fluctuations in its course and the ultimate recovery which sometimes occurs, if the œdema subsides and bone changes become arrested. But it is probable that simple œdema is not the

Fig. 79.—Caries of Spine (diagrammatic). Compression of cord by a carious abscess in the vertebral canal, at the region of the carious vertebra and external to the dura mater (the dark mass in the figure).

entire explanation; in many cases there is probably an inflammatory collateral œdema which is produced by some toxic substance. In consequence of œdema of long standing, degeneration of the nerve elements and softening occur. According to some pathologists, in certain cases,

owing to the compression, there is anæmia or ischæmia in the area of distribution of blood vessels within the cord, and this may lead to necrosis of the cord substance.

In some cases there is an inflammatory process which extends from the meninges to the cord, and plays a part in the degeneration of nerve elements.

The cord symptoms may therefore be due to (1) localised œdema, (2) to softening, as a result of œdema of long duration, or (3) to true inflammatory changes (myelitis).

Paraplegia due to compression myelitis may be produced *experimentally* in animals by injecting wax into the vertebral canal. The

Fig. 80.—Caries of Spine (diagrammatic). Compression of cord by projecting bone at the seat of the caries.

changes in the cord are similar to those found in compression myelitis in man. One cause of the compression myelitis is œdema of the spinal cord produced by obstruction to the flow of blood and lymph; but even in animals the cord changes may be marked when the obstruction to the blood and lymph circulation is slight. By placing *tuberculous* material on to the dura mater in experiments on animals, tuberculosis of the epidural tissue can be produced, and in course of time compression myelitis develops. In a minority of cases there is an extension of tubercular disease through the dura mater to the spinal cord; but in most cases this does not occur, and there is simply localised œdema of the cord with secondary degeneration of the nerve elements. In some cases the cord changes are due to compression, but in others the tubercular deposit in the dura mater is not sufficient to cause any considerable diminution of the space in the vertebral canal (*see* Schmaus and Saeki).

Symptoms.—The symptoms in compression myelitis produced by caries of the spine may be divided into three groups: (1) Those due to bone disease, (2) those due to affection of the spinal nerve roots by the diseased process at the seat of the caries, (3) those due to affection

of the spinal cord. In addition there is sometimes a little elevation of temperature; but often it is normal. Wasting and signs of tuberculosis affecting other parts of the body can be frequently detected.

In some cases of spinal caries only bone symptoms, or bone and nerve-root symptoms are present, and the cord is never affected: in other cases these symptoms are associated with symptoms of affection of the spinal cord.

1. *Bone symptoms.*—These consist of pains in the back, at the seat of the caries. The pain is increased by movement, and especially by pressure on the vertebral column. These symptoms are localised to the region of one or two adjacent vertebral spines; the region of the pain is always the same; and it is deep rather than superficial pressure which causes pain. On attempting to move the spinous process of the affected vertebra laterally, pain is felt. Pressure on the head or shoulders causes pain at the diseased region. When a sponge which has been soaked in hot water, or the kathode pole of the galvanic battery, is applied to various parts of the spine, pain is felt at the region of the caries. The patient holds the back rigidly, as if in a muscular splint. When he attempts to pick up any object from the floor, he does not bend the back, but holds it rigidly, and bends the knees and hips until his hands reach the floor. In cervical caries, the head is held rigidly (as described subsequently).

Deformity of the spine develops later than the pain. At the seat of the disease, a vertebral spine becomes prominent, or two or more adjacent spines may project, causing a distinct angular curvature. It is important to remember that sometimes no deformity or projecting vertebral spine can be detected.

An abscess may form at the seat of the caries, and the pus may gravitate to other parts of the body. Cervical caries may cause a retro-pharyngeal abscess or an abscess in the neck. Pus from a dorsal caries may pass into the mediastinum, or downwards into the sheath of the psoas muscle, and cause an abscess in the groin. In other cases, the abscess comes to the surface in the back.

2. *Nerve root symptoms* are marked in a few cases; usually they are only of moderate severity; often they are absent. They consist of pain in the distribution of the affected nerve roots, followed by hyperæsthesia, and later by anæsthesia. Sometimes there are spots of anæsthesia, or there is anæsthesia with pain (anæsthesia dolorosa), and herpes zoster occasionally develops in the distribution of the affected nerve roots. When the cervical or lumbar roots are affected, there may be weakness and atrophy of the muscles supplied by these nerves. In dorsal caries, the root symptoms consist chiefly of girdle pain, or of a zone of hyperæsthesia round the trunk.

3. *The spinal cord symptoms* are usually gradual in onset; but sometimes they are acute. They are those of a transverse lesion of the cord (resembling transverse myelitis). In most cases the paralysis (para-

plegia) is incomplete. Motor symptoms are more marked than sensory, and the latter may for a time be absent or slight. All forms of sensation may be affected in the severe case; there is then loss of sensation to tactile impressions, pain, and temperature, and loss of the muscular sense in the paralysed limbs: also, the patient is unable to feel the vibrations of a tuning fork placed in contact with bony points in the anæsthetic region.

But in other cases, the anæsthesia is partial, and one form of sensation is chiefly affected. Fickler found tactile sensation most frequently diminished; sensation to temperature was often diminished; whilst sensation to pain was diminished more rarely; and the sense of position was least frequently affected. I have sometimes found in early cases that the vibrating sensation (caused by the vibrating tuning fork) was lost when all other forms of sensation were present (*see* p. 80).

Bladder and rectal symptoms develop as in transverse myelitis, but they are absent in some cases and in others they do not appear until a late period.

The most common cord symptoms are those of spastic paresis with increased deep reflexes, ankle-clonus, and the extensor type of plantar reflex. When the motor weakness advances to complete paraplegia, there is usually anæsthesia with bladder symptoms. Unilateral paralysis (Brown-Séquard's paralysis) is very rare.

The peculiarities of the symptoms, in caries of various regions of the back, require to be briefly mentioned.

Dorsal caries is the most common form of the disease. The bone symptoms (local pain, tenderness and projection of one or more vertebral spines, etc.) are in the dorsal part of the vertebral column. An abscess connected with the caries may find its way into the sheath of the psoas muscle, and point at the region of Poupart's ligament. The root symptoms consist of a girdle sensation or pain around the chest or abdomen. The cord symptoms consist of paresis of the legs or paraplegia; usually the legs become rigid at an early period; the knee-jerks are increased; ankle-clonus is present; the superficial reflexes are increased and the plantar reflexes are of the extensor type (Babinski's reflex). The sensory symptoms are usually much less than the motor, and consist in formication, tingling, dull pain, hyperæsthesia, and later impaired sensation and anæsthesia (of the forms already described). The bladder and rectum are often, but not always, paralysed (as in dorsal myelitis). Bed-sores are often absent; when present they are usually slight. There is no atrophy of the leg muscles, and no reaction of degeneration. Occasionally the legs are flaccid and the reflexes lost, when the transverse lesion is complete.

In caries of the lower *cervical* and highest dorsal vertebræ, the bone symptoms are at the lower part of the neck; sometimes a projection is absent; but there is often considerable thickening of the structure in the neighbourhood of the vertebræ.

In caries of the lower cervical region affecting the 8th cervical and 1st dorsal nerve roots, there is pain with disturbance of sensation in the distribution of these roots along the inner side of the arm and hand (*see* p. 98). Also, there is weakness with atrophy of the small muscles of the hands, or of the muscles of the hands and forearm; and oculo-pupillary symptoms (contraction of the pupils and diminution of the palpebral fissures) are sometimes present. If the 5th and 6th cervical nerve roots are implicated there is atrophy and paralysis of the deltoid, biceps, brachialis anticus, and supinator longus, and the anæsthesia extends to the distribution of these roots at the outer side of the arm.

The paralysis and atrophy of arm muscles may be produced both by affection of the cervical nerve roots and also by cord changes in the spinal segment, from which the root arises. The atrophied and paralysed muscles often present the reaction of degeneration on electrical examination.

The intercostal muscles are paralysed, there is paralysis or paresis of the legs, and of the bladder and rectum, as in dorsal caries. The legs are usually spastic with increase of the reflexes as already described under dorsal caries. (Whilst the arm muscles present atrophic paralysis.) The sensory affections are of the same nature as in dorsal caries. The upper limit of anæsthesia may be at the upper part of the thorax, and in addition the anæsthesia may extend down the inner side of the arm and hand, or over a larger area according to the level of the lesion.

In caries of the *upper cervical* region there is pain at the back of the neck and posterior part of the head, stiffness in the neck, a feeling of weight in the head, and deformity of the neck (prominent spine, etc.) at the upper part. All four limbs may be weak and spastic. The sensory impairment extends higher (the upper limit depending on the seat of the lesion). The phrenic nerve is very liable to be affected and respiratory paralysis often occurs, both diaphragm and intercostal muscles being paralysed.

In caries of the *lowest dorsal and first lumbar* vertebræ, the bone symptoms are in the region of the vertebræ affected. An abscess in connexion with the diseased region usually extends into the sheath of the psoas muscle, and points below Poupart's ligament, or in the lumbar region of the back. The root symptoms consist of pain in the legs, especially in the region of the sciatic nerves, or of pain in the region of the bladder, rectum, and perineum. The legs are paralysed and flaccid, the affected muscles atrophy and present the reaction of degeneration. The knee-jerks and reflexes in the legs are absent, the bladder and rectum are paralysed, and there is incontinence of urine from the first. Bed-sores usually develop over the sacrum, and often become very extensive.

If only the conus terminalis is affected, the symptoms will correspond to those described under the account of conus lesion along with bone symptoms in the region of the vertebræ diseased.

In caries of the *first and second cervical vertebræ*, the atlas, the odontoid process of the axis, and the atlanto-occipital joint are liable to be affected. The patient suffers from headache and pain in the neck. He holds the neck and head rigidly, and avoids movements and rotation of the head. Rotation of the head and nodding movements cause pain. On raising the head from the pillow, or other support on which it is resting, he holds it on each side with his hands. Slight pressure on the head causes pain. Frequently there is unilateral or bilateral occipital neuralgia, and later anæsthesia in the area of distribution of the occipital or upper cervical nerves develops. Dizziness and nystagmus may be present. Pressure on the medulla may occur and symptoms of bulbar paralysis are then produced. Sometimes symptoms of a myelitis at the highest cervical region develop. The arms may be affected first, and then the legs (though the reverse order of development of these paralytic symptoms is sometimes noted). The paralysed arms are not atrophied. Other symptoms are difficulty of swallowing, vomiting, slowness of the pulse, and dyspnœa.

Paralysis of the spinal accessory and sometimes of the hypoglossal nerves occasionally occur.

Retro-pharyngeal abscess, or abscess in the neck, may develop. Sudden death occasionally results from fracture of the odontoid process and sudden dislocation. When only the joint between the odontoid process and the atlas is diseased, the patient cannot rotate the head, or can only do so with difficulty, whilst the nodding movement is not affected. Occasionally the head is drawn to one side, as in torticollis.

The *course* of spinal caries with compression myelitis is chronic. In most cases the spinal symptoms develop slowly. In rare cases there is a rather sudden onset of the paralysis, owing to spinal curvature occurring suddenly through breaking down of a carious vertebra. Often the symptoms of affection of the spinal cord, and the signs of vertebral disease (curvature or prominent vertebral spine) develop about the same time. But occasionally the spinal curvature is present for a long period, or even many years before the symptoms of affection of the spinal cord develop. Thus, in rare cases, an angular curvature has developed in childhood and spinal symptoms have not appeared until adult life. In other cases the symptoms of compression myelitis develop before any spinal curvature or prominence of vertebral spines can be detected, and the symptoms of vertebral disease may remain very slight.

After the rupture of an abscess connected with a carious vertebra the paralysis may disappear; but the paralysis may persist although the carious process heals; in other cases the caries continues though the paralysis disappears.

When the termination is fatal, it is usually due to the development of complications such as bed-sores, cystitis, pyelonephritis, pyæmic conditions, and general tuberculosis. Often, however, the paralysis is present for a long period (even years) before death occurs from the compli-

cations mentioned. Sometimes almost complete recovery occurs, only slight paresis remaining. In a minority of cases, the recovery is complete, and with careful treatment the proportion of such cases ought to be much greater.

As already mentioned the observations of Schmaus on the pathology of compression myelitis have shown, that at first the cause of the paralysis, in many cases, is œdema and lymph stasis at one part of the cord, and that a long period may elapse before incurable softening, inflammatory degeneration, or sclerotic changes occur.

Prognosis.—Unfavourable signs are paralysis of the bladder, cystitis, bed-sores and paralysis of the intercostals. The more marked the spinal symptoms, the worse the prognosis. When the reflexes are absent and the paralysis flaccid, the prognosis is worse than when the legs are spastic and the reflexes increased. The prognosis is worse when the disease is in the cervical region (especially upper cervical) than in the dorsal part. It is worse in adult and elderly people than in young persons and children. The presence of general tubercular disease is, of course, a most unfavourable sign.

Diagnosis.—The *early recogniton* of caries of the spine, and especially its recognition as the cause of compression myelitis, is of *great importance.* By suitable treatment, especially by prolonged rest in bed, at the early period of vertebral caries, recovery may be obtained without the spinal cord being affected. And in those cases in which compression myelitis is developing, or has already developed, the early recognition of spinal caries as its cause, is of importance; because by suitable treatment at this stage, recovery may be obtained more readily than at a later stage.

The diagnosis of compression myelitis due to caries of the spine is usually easy, but in some cases it is very difficult, especially at the early stage, or when signs of spinal deformity and bone disease are slight. In many cases, in which there ought to be no difficulty, the cause of the paralysis is not recognized, because the spine is not examined. Hence it is a good practice to examine the back for signs of a prominent vertebral spine or other indication of caries in all cases of spinal cord disease, more especially when spastic paraplegia with increase of the reflexes and a girdle sensation have developed gradually, since symptoms of this nature are so very often due to caries.

A diagnosis of compression myelitis from spinal caries is justified, in a case presenting symptoms of a transverse lesion of the cord, when a definite projecting vertebral spine can be detected, and root symptoms have developed, providing there be no indications of spinal tumour, hydatid, or syphilitic disease. Local signs of vertebral disease distinguish the compression myelitis of caries from transverse myelitis unassociated with caries.

Local pain and tenderness on percussion, limited to one or two vertebral spines, are important diagnostic signs, and the testing for local

pain by means of a hot sponge is of service. Angular curvature, or a prominent vertebral spine, is best marked in the dorsal region ; in the lumbar and cervical region it is often slight or indefinite. In the cervical region there is often thickening of the tissue at the back of the neck in the region of the disease, owing to infiltration of the periosteum and surrounding soft tissues.

Important evidence in favour of caries in cases of doubtful diagnosis, is the coincidence at one region of a definite prominence with deep tenderness. If an abscess should be present at one of the regions at which abscesses connected with spinal caries tend to point, this is a valuable aid to diagnosis.

The fact that compression of the cord or nerve roots sometimes occurs before any spinal deformity can be detected, makes the diagnosis in certain cases very difficult. In such cases, localised pain and deep tenderness, if well marked, are of great importance. The course of the symptoms is of value : at first there is persistent pain in the back, followed by gradually increasing paralysis with girdle pains, and preponderance of paralysis or paresis over anæsthesia or impaired sensation. Another important feature is the peculiar stiffness or rigidity of the back—the want of mobility of the back noticed during various movements (sitting down, getting up, turning round, walking, or stooping to pick up anything from the floor). The patient holds his back rigidly in a kind of muscular splint. When the cervical spine is affected, the head is held rigidly (as already described). The presence of tubercular disease in other parts of the body is a sign of great value. (In several cases of paraplegia from caries in which there was no prominent vertebral spine, the author has found that the localised pain in the back was increased on sitting up, but relieved on lying down.)

In all cases repeated examination for a definite prominent vertebral spine is, of course, most important. To be of diagnostic value, the prominence of the vertebral spine should be distinct, since a very slight prominence of one or two spines can be sometimes detected in health. Further, in rare cases of compression myelitis due to spinal caries, the curvature is chiefly lateral ; but it is to be remembered that most cases of lateral curvature of the spine are not due to caries of the vertebræ.

Examination with the *Röntgen* rays is great diagnostic value in many, but not in all, cases. When prominence of a vertebral spine is absent or indefinite, the Röntgen rays may show the presence of caries. In radiograms of such cases the clear interval, which ought to be seen corresponding to the discs between the vertebræ, may have disappeared, owing to fusion of the bodies of two vertebræ. The bodies of one or more vertebræ may also appear altered in shape, and irregular dark shadows may be noted corresponding to masses of caseous exudation. It is in cervical caries that the X rays are most likely to give definite results.

In cases of spinal irritation, and in neurasthenia or hysteria the question of caries may arise; but in the former affections there are several tender spots, or the tenderness is over a considerable area of the spine, and not localised to one or two vertebræ as in caries; also in neurasthenia the tenderness is both superficial and deep, and root pains are absent. An important sign in favour of caries would be the rigid position in which the spine is held during various movements. If, in addition, a prominent vertebral spine is present, caries is indicated more clearly. In the diagnosis of the paralysis due to compression myelitis from hysterical paralysis the points already mentioned are of service (*see* p. 136), especially the Babinski type of plantar reflex. If X ray photography should reveal a definite shadow, this fact would be against neurasthenia or hysteria being the sole cause of the symptoms.

When the vertebral symptoms of caries are associated with those of a compression myelitis presenting the features of an organic paralytic affection, the condition cannot be mistaken for hysteria, unless the examination be most superficial and careless. It is to be remembered that patients (especially females) suffering from caries of the spine may have hysterical symptoms also: and unfortunately, in such cases a wrong diagnosis is sometimes given, and all the symptoms attributed to hysteria.

In cervical caries when the arms are wasted and paralysed, the symptoms may resemble those of amyotrophic lateral sclerosis or progressive muscular atrophy. But the pain and sensory disturbances in the former case with the signs of bone affection are diagnostic.

The diagnosis from aneurism and tumour compressing the cord is considered p. 159.

Treatment.—For the successful treatment of compression myelitis from spinal caries, early diagnosis is of the greatest importance.

The two chief principles of the medical treatment are: (1) to improve the general condition; (2) to secure perfect rest to the vertebral column. The latter is best secured by rest in bed, on the back, for months. An abundant supply of food, including a large amount of fatty substances (cream, milk, and cod-liver oil) is desirable. If possible, the bed or couch should be wheeled out into the open air when the weather is suitable, and the open air treatment for tubercular disease should be carried out. General tonics such as iodide of iron or other iron preparations are often given, and creosote, calcium sulphide and calcium phosphate have been much recommended. It is most important to prevent the formation of bed-sores by perfect cleanliness, by washing the skin of the sacral region with lotions (*see* p. 137), and by the use of a water cushion or water bed. When the bladder is paralysed, the use of the catheter will probably be necessary; and great care will then be required to prevent cystitis (*see* p. 138). If cystitis should develop, the use of antiseptic lotions and the administration of urotropin may be necessary as in cases of myelitis (*see* p. 138).

When the leg muscles become spastic, galvanism and massage are not of service; but when flaccid, both may be useful (*see* p. 139). For the spasmodic condition of the legs Starr recommends 10 grains of sodium bromide every two hours.

By rest in bed, prolonged for months, and by general tonic treatment, excellent results can often be obtained, many cases being cured. In cervical caries it is necessary to fix the head and neck in bed by long sand bags applied to each side of the head.

In some cases persistent extension of the spine, in the recumbent posture, has been followed by good results. A weight of four to twelve pounds is attached to a band passing round the patient's waist above the hips, and the weight is allowed to hang over the foot of the bed; the upper part of the spine is fixed to the head of the bed, by bands passing under the arms in disease of the dorsal vertebra, or by a band attached to the patient's head under the chin, in cervical caries. In this way continuous extension of the spine can be produced.

Another way of obtaining extension of the spine is by tilting the bed. Chiene recommends that in cervical caries a weight, hanging over a pulley at the head of the bed, should be attached to the patient's head, and the head of the bed raised, so that the body acts as a counter extending force; in lumbar caries the foot of the bed should be raised and the weight attached to both legs, the body acting as the counter extending force. In dorsal cases, double extension should be employed from the head and the legs (the bed being horizontal). The forcible extension of the spine recommended by Calot is not advisable; in some cases it is distinctly dangerous.

After a prolonged rest, if the disease appears to have become quiescent or arrested, suspension and the application of a plaster-of-Paris jacket may be employed, and the patient is allowed to walk about again. This is often followed by improvement. In cervical caries a "jury-mast" to support the weight of the head is believed to be of service after a prolonged rest in bed.

When there is great return of power in the legs, but walking is prevented by the contraction of the calf muscles, tenotomy (division of the tendo Achillis) may be of great service. The foot may then be brought into its normal position and the patient enabled to walk. When an abscess connected with vertebral caries can be detected in the groin, back, or other part, it will require to be opened with antiseptic precaution.

In certain cases other operative treatment, laminectomy and removal of diseased tissue, has been of service. But it is only suitable in a few cases, and as the operation is not without risk, it ought not to be employed until the treatment by prolonged rest has been tried. It is contra-indicated when signs of rapidly advancing general tuberculosis are present, and also in chronic cases in which the kidneys, liver, or other organs are diseased.

The removal of inflammatory products which have collected outside

the dura mater may give relief ; and if the cause of compression cannot be taken away, the removal of one or more vertebral arches sometimes relieves pressure. Sir William Gowers and Dr. J. Taylor think "an operation is suggested whenever a sudden increase in the curvature or severe root pains coincide with the acute onset of paralysis." When these symptoms are absent the rapid onset is probably due to acute myelitis, and then operation would be useless. In cases of disease in the upper part of the cord, causing danger to life from respiratory failure, operation may be necessary.

Oppenheim thinks operation is indicated (1) in the rare cases of caries of the vertebral arch, when the disease does not subside by medical treatment ; (2) also when the opening of an abscess leads directly to the seat of the disease of the body of a vertebra ; and (3) when the paralysis persists after long duration of the affection, and after apparent healing of the vertebral disease.

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Arthritis Deformans and Allied Chronic Diseases of the Vertebral Articulations (Vertebral Ankylosis, Spondylose Rhizomélique).

Various joint affections, in rare instances, implicate the vertebral articulations. The most common of these affections is arthritis deformans. The condition may lead to complete ankylosis of the vertebral articulations and rigidity of the vertebral column, with a peculiar stooping attitude. There is pain in the region of the vertebral column, which is increased by movement. This pain is often due to the joint disease ; but in other cases the nerve roots are compressed by new formation of bony tissue. The latter condition causes radiating pain in the arms, legs, and trunk and slight muscular atrophy may be produced. In extremely rare instances the spinal cord has been compressed. The diagnosis is based on the evidence of arthritis in other joints, the rigidity (ankylosis) of the vertebral column (even under chloroform), and the nerve root pains. Sometimes new bone formation can be felt in the back. The treatment is that of arthritis deformans. Salol has been especially recommended by Oppenheim.

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SPINAL TUMOURS

Tumour affecting the spinal cord is a very rare disease. In the records of 35,000 post-mortem examinations at the Vienna General Hospital, Schlesinger found only 151 cases in which a tumour was present in the vertebral column, spinal meninges or spinal cord. In only 104 of these cases was the spinal cord affected, directly or indirectly.

Tumour of the brain is much more frequently met with than spinal tumour.

The tumour growths affecting the spinal cord may be divided into the following groups, according to the structures in which they commence.

1. Vertebral.
2. Meningeal extra-dural (outside the spinal dura mater).
3. Meningeal intra-dural (inside the spinal dura mater).

} Extra-medullary spinal tumours.

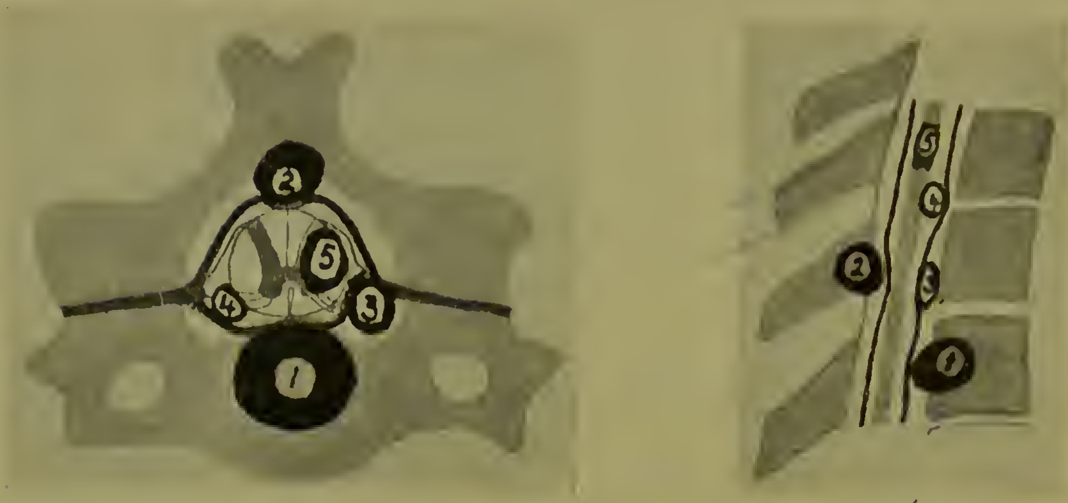
4. Intra-medullary spinal tumours.

Growths commencing in the first three positions cause "compression myelitis." Intra-medullary spinal tumours destroy the cord substance and produce symptoms of a transverse lesion.

The following table from Schlesinger shows the nature and position of 400 tumours of the spinal cord :—

Variety.	Intra-dural.			Extra-dural.		Single.	Multiple.	Total.
	Medullary.	Meningeal.	Both.	Meningeal.	Non-Meningeal.			
Sarcoma	14	53	9	17	11	80	27	107
Tubercle	62	—	—	2	—	55	9	64
Echinococcus	—	5	—	39	—	8	36	44
Fibroma	—	20	2	5	—	15	18	33
Gumma	7	4	15	2	—	19	9	28
Glioma	20	—	—	—	—	20	—	20
Psammoma	—	18	—	—	—	18	—	18
Myxoma	—	7	—	4	—	11	—	11
Lipoma	1	8	—	—	1	8	3	11
Cysticercus	2	5	—	—	1	4	4	8
Glio-sarcoma	—	3	4	—	—	—	7	7
Endothelioma	—	5	—	1	—	4	2	6
Melano-sarcoma	1	—	3	—	—	1	3	4
Neuroma	4	—	—	—	—	3	1	4
Lymphangioma	—	1	—	1	—	1	1	2
Cysts	—	1	—	1	—	1	1	2
Cholesteatoma	1	—	—	—	—	1	—	1
Uncertain	13	12	2	3	—	24	6	30
	125	142	35	75	13	273	127	400

Meningeal tumours are more common than intra-medullary, and tumours of the vertebræ, implicating the spinal cord, are nearly twice as numerous as all other forms of meningeal and intra-medullary spinal tumours together. (In the following account of the pathological anatomy the author is much indebted to Schlesinger's thorough description and analysis of cases.)



A. B.

FIG. 81.—Diagrams to show positions at which Spinal Tumours may develop.
A = Transverse section of a cervical vertebra and of the spinal cord surrounded by the dura mater. Extending on each side is a spinal nerve, indicated by a deep black line.
B = Longitudinal section of vertebræ and of the spinal cord with dura mater.
Positions of tumour :—(1) Vertebral (in body). (2) Vertebral (in arch) (3) Extra-dural (meningeal). (4) Intra-dural (meningeal). (5) Intra-medullary.

VERTEBRAL TUMOURS

Malignant vertebral tumours are more common than benign (10 to 1).

Malignant tumours may be primary or secondary. The chief primary growths are sarcoma and myeloma ; the secondary, carcinoma, sarcoma, and lympho-sarcoma. Primary carcinoma of the vertebræ does not occur.

Primary sarcoma is very often multiple (in about half the cases) and affects several vertebræ. It may develop in the body of the vertebra or from the periosteum ; or in rare cases from a transverse or spinous process. The growth extends beyond the limits of the bone, and compresses the spinal cord ; it may invade, but usually does not perforate, the dura mater.

Secondary sarcoma is probably a little less frequent than the primary. The growth is often multiple, and it may be nodular or diffused. The bodies of the vertebræ are most often affected and adjacent parts, muscles and membranes, etc., may be invaded. The sarcoma may be secondary to growths in other bones or other parts of the body (metastatic) ; or it may extend to the vertebræ from adjacent tissues.

Lympho-sarcoma may affect the vertebral column in a diffuse manner, without causing any alteration in form. In the cases on record of the rare form of green coloured tumour, known as chloroma, the vertebræ have often been affected.

Multiple myeloma or myelo-sarcoma has affected the vertebral column in several of the cases recorded, and has led to compression of the cord.

Carcinoma of the vertebræ is always metastatic or secondary. Often there is extensive carcinomatous infiltration in the vertebræ and many vertebræ may be affected, whilst the primary growth elsewhere may be very small and very easily overlooked at the autopsy. In the metastatic form, the primary growth is in the breast, thyroid, uterus, bronchus, stomach or prostate : in the secondary form, the growth commences in the œsophagus or stomach, and extends to the vertebræ.

According to the statistics of the Vienna General Hospital medullary carcinoma is the most frequent form; then follow in diminishing frequency epithelial carcinoma, scirrhus, adeno-carcinoma. The growth scarcely ever extends through the dura mater except in the region of the cauda equina.

The benign vertebral tumours may not produce any affection of the spinal cord, but cases are on record in which the cord has been compressed or affected by the following benign tumours :—Exostosis, osteoma, angioma, chrondroma, myxoma. Benign growths seldom develop from the dorsal side of the vertebræ, usually they grow from the ventral side, or from the anterior or inner side of the vertebral arches.

In tumour of the spinal column the vertebræ are often eroded and softened by the growth, and spinal curvature is produced, chiefly forwards, rarely laterally. The height of the vertebral column is diminished and fracture and dislocation of the vertebræ may occur. Often several vertebræ, occasionally many, are invaded by the growth. The tumour may extend to the surrounding tissue outside the bone, and form a subcutaneous swelling in the back. The nerve roots are usually compressed by the growth at the inter-vertebral foramina. The tumour extends between the dura mater and bone of the vertebral canal, but it does not usually penetrate to the inner side of the dura, also it never invades the cord. At first the paralytic symptoms and changes in the cord are often due to œdema through compression of spinal blood vessels and lymphatics. Afterwards softening may occur. In some cases acute transverse myelitis occurs at the region of the vertebral tumour. In other cases there is mechanical pressure of the growth on the cord. Occasionally the growth causes softening by obstructing a spinal artery through pressure, or by producing obliterative endarteritis.

Symptoms.—(1) *Vertebral.* There is usually, but not always, pain and tenderness in the spine at the region of the growth. It is generally very severe and boring in character ; it is increased by movement, and by pressure or percussion. Curvature of the spine sometimes occurs at the seat of the growth, but it is not so sharp as in caries, and is more rounded than angular. The spinal deformity may be kyphosis, scoliosis, or a lateral displacement. A tumour sometimes can be felt and seen posteriorly on one side of the spinous processes at the seat of the lesion, but often it is late in development. There may be rigidity of the spine at the region of the tumour, and, according to Schlesinger, in rare cases local œdema.

Owing to the sinking down of the diseased vertebræ the height of the body may appear to be diminished.

(2) *Nerve root symptoms.* By pressure on the spinal nerve roots, at the seat of the tumour, pain is caused, which radiates in the course of the nerves compressed. The pain is usually exceedingly severe, and is much greater than in caries. It is sharp and stabbing ; at the early stage it is intermittent ; later it becomes constant, with paroxysms. The latter are produced by movement, and jarring or shaking of the body. The seat of the radiating pain is in the legs, arms, or trunk according to the region of the vertebral tumour. Tumour of the lumbar vertebræ may cause double sciatica. Hyperæsthesia often occurs in the course of the nerves compressed ; later anæsthesia or patches of anæsthesia may develop ; and occasionally there is herpes zoster. By pressure on the motor nerve roots, spasm of muscles and later paralysis may be caused. The nerve root symptoms may be unilateral at first, then they become bilateral. The root symptoms may precede other symptoms of cord affection for a long period, often for months.

(3) *Cord Symptoms.* These closely resemble those of compression myelitis from caries of the spine, and for a description the section on that disease may be referred to. But the paralysis often develops much more rapidly than in caries. This is owing to the cord changes being often caused by acute myelitis at the region of the growth. The paralysis of the legs may become complete in twenty-four hours, but in other cases the onset of the paralysis is very gradual. According to Gowers, there is sometimes retention of urine for several days previous to the paraplegia. Sensation is more frequently lost in compression from vertebral tumour than in spinal caries. Brown-Séquard paralysis is occasionally observed at first, when the cord is becoming affected.

The paralysis in vertebral tumour is atrophic in the muscles at the level of the lesion ; but usually spastic in parts below the lesion.

Schlesinger has described thrombosis of the large veins of the legs, and sudden death through embolism of the pulmonary artery, in cases of carcinoma of the vertebræ.

In multiple myeloma albumose[1] may be present in the urine ; and elevation of temperature and anæmia may occur.

The causes of death are the same as those in myelitis from caries —septic infection from cystitis or bed-sores, respiratory paralysis, and other complications, and the cachexia and complications of malignant disease.

In vertebral carcinoma death usually occurs in from twelve to eighteen months ; in sarcoma the duration may be longer.

Diagnosis.—In a case of paraplegia the diagnosis of compression myelitis due to vertebral tumour is chiefly based on : (1) evidence of primary growth in some other part of the body ; (2) spinal curva-

[1] The tests for albumose are well described by Bradshaw, *British Medical Journal*, November 25, 1906.

ture, with vertebral pain increased by movement or pressure; (3) evidence of a tumour at one part of the spine, or close to the spinal column in the back, or in the posterior triangle of the neck; (4) the presence of growths in the superficial lymphatic glands (especially in the groin or neck); (5) the very great severity of root pains. (Severe pains in the district of one or several adjacent nerve roots are suggestive of vertebral growth, if a tumour has been previously removed from some part of the body, especially from the breast.) (6) Signs of cachexia, and absence of indications of tubercular disease, are also of some value. (7) Leyden and Grunmach have shown that a vertebral tumour may be sometimes recognised by the X rays. In one of their cases the X ray photograph gave clear evidence of a carcinoma of the sixth and seventh cervical vertebræ, which was secondary to a cancer of the breast. In another case the X rays gave evidence of a sarcoma of the lumbar vertebræ, secondary to sarcoma of the thigh.

In the diagnosis from caries the age is of some value. In early life caries is more common than tumour; in the second half of life, according to Gowers, the frequency of the two affections is about the same. In tumour the pain is much more severe and increased more by movement. The presence of a tumour in the back close to the spine, or of a tumour (or evidence of the removal thereof) in some other part of the body would point to vertebral tumour. The presence of tubercular disease of the lungs or other organs would point to caries. Spinal curvature caused by tumour is less angular than that due to caries. Abscess formation is in favour of caries.

At an early period, when only root symptoms are present, mistakes in diagnosis are very liable to occur. (In one case seen by the author a diagnosis of gall stones had been made at the early period of the disease; in another case renal colic had been diagnosed.)

MENINGEAL AND INTRA-MEDULLARY SPINAL TUMOURS.

Pathology.[1]—Intra-dural tumours are met with more frequently than extra-dural. Intra-medullary tumours occur chiefly in the cervical and lumbar regions; extra-medullary (meningeal) chiefly in the dorsal region. (Cystic tumours will be considered, p. 179.)

1. *Extra-dural* (meningeal) tumours may arise from the dura mater, from the tissues between the dura mater and the vertebræ, or they may grow through the inter-vertebral foramina from the surrounding tissues. The chief forms of extra-dural tumour are sarcoma, hydatid cyst, fibroma, lipoma, myxoma, fibro-sarcoma, myxo-sarcoma, and lipo-sarcoma, lymphangioma, and metastatic sarcoma and carcinoma. Extra-dural spinal tumour may be due to the extension of growths from the vertebræ or their periosteum—carcinoma, sarcoma, enchondroma, exostosis (these have been already considered).

[1] In the following account of the pathology the author is much indebted to Schlesinger's analysis and description.

2. *Intra-dural* (meningeal) tumours may arise from the inner surface of the dura-mater, from the arachnoid, or from the pia mater. The chief forms of tumour arising in this region are sarcoma, fibroma, psammoma, myxoma, lipoma and myo-lipoma, endothelioma, syphilitic gumma, fibro-sarcoma, fibro-myxoma, tubercular tumours, angioma, cysticercus, and, very rarely, hydatid cyst. Neuromata and neuro-fibromata and sarcomata are occasionally found on the nerve roots, and pressure on the cord may be caused by these growths. It is very rarely that the intra-dural tissue is affected by cancer, either by extension from the outside or by metastatic tumours.

3. Growths commencing within the cord, *intra-medullary*, are more rare than the meningeal tumours. They may arise from the pia mater or the cord substance, and especially from the tissue around the central canal. The chief forms are tubercular tumour, glioma, sarcoma, syphilitic gumma, myxoma, myxo-sarcoma, glio-sarcoma, fibro-glioma, neuroma. Gliomata often begin in the tissue around the central canal, gummata from the inner side of the pia mater, tubercular growths from the cord substance. Metastatic tumours within the cord are extremely rare.

Tumours affecting the cauda equina are chiefly fibroma, neuro-fibroma, fibro-sarcoma, sarcoma, syphilitic gumma, glioma, and lymphangioma,

Forms of Tumour Growth.—*Carcinoma* is rare and is always secondary. It may occupy the subdural space and spread to the dura, but does not break through it, except at the region of the cauda equina. In the cord itself metastatic carcinomatous growths have been met with occasionally in cases of universal carcinoma.

Metastatic and *secondary sarcoma.* Whilst carcinoma very rarely breaks through the dura matter, secondary or metastatic sarcoma not infrequently breaks through all the meninges and invades the cord.

Primary sarcoma may be solitary or multiple. Solitary sarcoma may commence in the cord, in the meninges, or in nerve roots. In the two forms last mentioned the growth may remain localised to the meninges or nerve roots, or it may extend to the cord.

Primary solitary sarcoma of the cord is very rare. It is met with chiefly in the cervical region. The structure may be that of a spindle-celled, round-celled, cysto- or angio-sarcoma.

Primary solitary sarcoma of the meninges and nerve roots is the most frequent form of meningeal tumour, and is usually either a fascicular or round-celled sarcoma. The fascicular sarcoma of the meninges and nerve roots grows slowly, causes no metastatic growths and usually does not invade the cord. It is usually hard, sharply limited, and surrounded by a capsule. The round-celled solitary sarcoma spares the cord for a long time and usually affects it only by compression. The tumour is sharply limited and often encapsuled. This form sometimes gives rise to metastatic growths.

In primary multiple sarcoma of the central nervous system the

meninges are extensively invaded; in half the cases the cord is not affected except by compression. The growth may invade the nervous system in a diffuse manner, and cases have been recorded in which the meninges were chiefly affected, the macroscopic appearances resembling meningitis.

Metastatic multiple glio-sarcoma of the spinal cord and meninges usually follows glio-sarcoma of the eye-ball.

Endothelioma may be solitary or multiple. The growths commence from the inner (or rarely from the outer) side of the dura mater or from the arachnoid, and may compress the cord.

Psammoma forms a solitary sharply defined growth which develops very slowly. It usually commences on the inner surface of the dura mater, and often compresses the cord markedly, but does not invade

A B

FIG. 82 (A and B).—Tumour of Dura Mater (Sarcoma), Extra-dural. Cervical Region. In Fig. A the dura mater had been divided longitudinally on the anterior surface and the two portions extended laterally. Fig. B shows the cord with dura mater seen from the side. At the upper part the dura mater had been cut away. The tumour is seen attached to the outer side of the dura mater, on its posterior aspect.

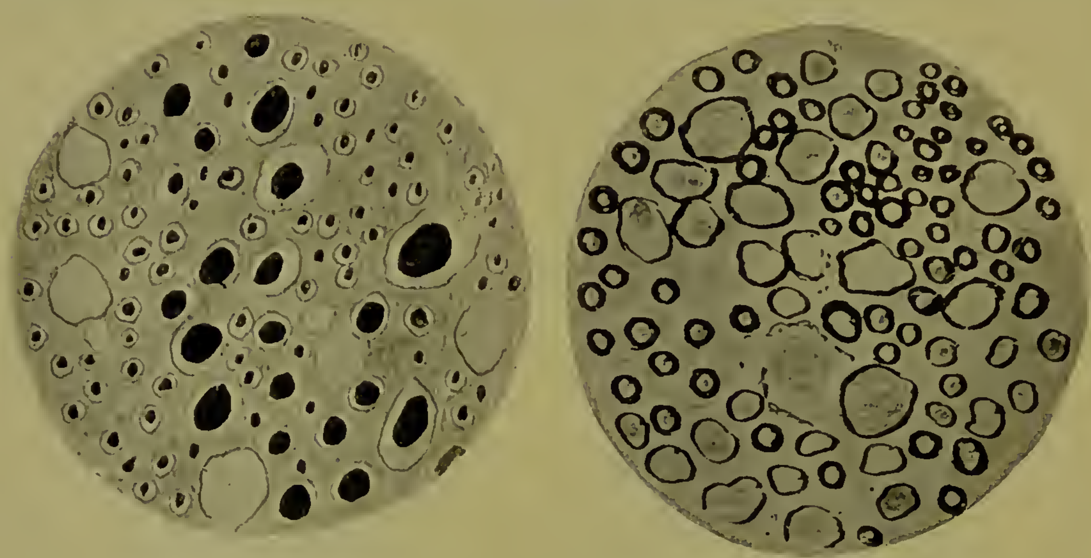
it. The structure consists of small spindle-formed cells which often have a concentric arrangement. The growth is frequently calcified. Spinal psammoma occurs chiefly in old women.

Solitary fibroma and neuro-fibroma are sharply defined growths. They are more frequently intra-dural than extra-dural, and arise from the dura mater and delicate membranes or from nerve roots. The solitary fibroma is not larger than a plum stone. It is firm and grows slowly, and usually affects individuals over thirty. In multiple fibroma

(neuro-fibroma) very often the peripheral nerves, the spinal nerve roots within or outside the dura, and the cauda equina are affected. Sometimes the tumour invades the cord.

Myxoma is usually solitary; more frequently it is intra-dural than extra-dural; it arises from the dura mater or delicate membranes. Most frequently it occurs in the dorsal region. Often the tumour is of a mixed form—myxo-sarcoma, myxo-lipoma etc.

Lipoma occurs in childhood, and may be associated with spina bifida. The growth may be extra- or intra-dural and may compress the cord. It is not infrequently multiple; the tumours are benign, and often the growth is of a mixed form—myo-lipoma. The lower end of the vertebral canal is most affected by the growth.



A B

FIG. 83. — Transverse section of the Spinal Cord. Compression Myelitis from meningeal tumour, showing swollen axis-cylinders and dilated myelin sheaths.
A = Formol and nitrate of silver stain, axis-cylinders deep black.
B = Weigert's stain. Axis-cylinders pale. Myelin sheaths deep black. Some normal fibres, others with swollen axis-cylinders and dilated myelin sheaths.

Other rare forms of spinal tumour are occasionally met with:—Adeno-sarcoma, teratoma, lymphangioma, cylindroma, angioma and cholesteatoma.

Tubercular tumours may occur in several forms in the meninges and in the cord, and often the extra- and intra-medullary forms are combined (*see* p. 386). With the exception of the tubercular or caseous external pachy-meningitis, associated with vertebral caries, tuberculosis of the cord itself is the most common form. It may occur as miliary tubercles, or as a mass of conglomerate tubercles. The first form is rare; a solitary or conglomerate tubercular mass is much more common, and is the most frequent form of intra-medullary tumour. The tumour may attain the size of a cherry stone. The form is roundish, and the centre is caseous. Often

the brain and spinal membranes are also affected by tubercular disease. Sometimes several tubercles are present in the grey matter at different levels.

The spinal tubercle is always secondary; the meninges are also affected in three-fourths of the cases, and tubercular caries is not infrequent. Solitary tubercle occurs most frequently at the lower end of the cord.

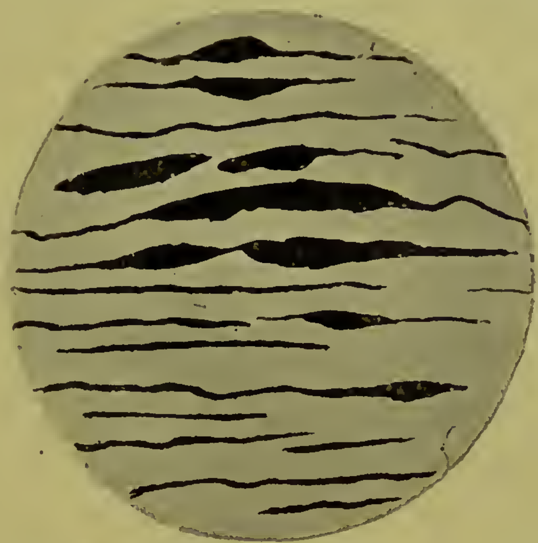


FIG. 84.—Compression Myelitis: longitudinal section of white matter: spindle-shaped swellings of axis-cylinders: formol and silver nitrate stain.

Gumma is described on page 394. It may be meningeal or intra-medullary.

A glioma is composed of cells and fibres resembling the neuroglia cells and fibres. The cells have numerous processes, which do not anastomose but form a network. The network of neuroglia fibres is absent in sarcomata. Gliomatous tumours are intra-medullary; they are limited to the cord substance; they do not invade adjacent structures, and do not give rise to metastatic growths: the meninges are not affected. The growths are often much elongated. The structure of the cord is often entirely destroyed by the growth, and cavities are sometimes present in the tumour which resemble those of syringo-myelia. Also gliomata are sometimes associated with syringo-myelia.

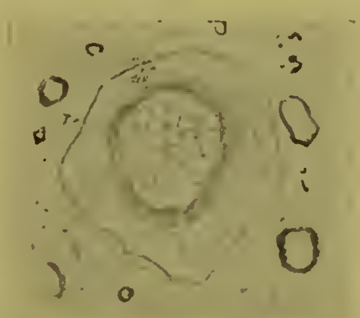


FIG. 85. — Blood vessel with dilated perivascular sheath. Compression myelitis from tumour.

Many of the cells of the gliomatous tumour resemble neuroglia cells, others are multinuclear.

Sometimes the tumours contain large cells somewhat resembling ganglion cells (neuroglioma ganglio-cellulare).[1]

General Pathology. — Spinal tumours, meningeal and intra-medullary, are often small, but sometimes they extend for a long distance in the vertical direction. Usually spinal tumours are single, occasionally they are multiple.

Sometimes, as in cases of sarcoma, a meningeal growth invades the cord substance, but usually meningeal tumours only affect the cord by pressure causing "compression myelitis." In some instances the compression myelitis is due to direct mechanical pressure of the growth

[1] See Zeigler's *Textbook of Pathology* and writer's paper in *Edinburgh Medical Journal*, July, 1902.

on the cord which is indented and diminished in diameter; but more frequently the growth impedes the circulation of blood and lymph at the seat of the lesion, by diminishing the space in the vertebral canal, and local œdema and later softening and degeneration of the cord follow (as in compression myelitis from caries). In other cases the tumour growth excites a localised transverse myelitis, or it causes softening by obstructing a spinal artery through direct pressure or by causing obliterative endarteritis.

The "compression myelitis" is followed by the usual ascending and descending degeneration.

In intra-medullary tumour there is often softening just around the growth.

As regards the **Etiology** of spinal tumour that of tubercle, gummata and parasitic cysts is the same in these lesions as in other parts of the body. The cause of other forms of tumour is not known. Injury may have played some part in the etiology in a few cases.

According to Schlesinger the most frequent forms of tumour at different ages are as follows:—

	Intra-medullary.	Extra-medullary.
Under 10 years . . .	Tubercle.	Lipoma or sarcoma.
From 10–20 ,, . . .	Tubercle and glioma.	Multiple and metastatic sarcoma and hydatid cyst.
,, 20–40 ,, . . .	Tubercle and glioma.	Sarcoma and hydatid.
,, 40–60 ,, . . .	Gumma and tubercle.	Solitary sarcoma, psammoma.

Symptoms.

I. *Meningeal tumours* (*extra- and intra-dural*).

Briefly stated the typical symptoms of meningeal tumour consist of—

(i) "*Root symptoms*" (unilateral then bilateral), followed in course of time by

(ii) *Cord symptoms*

(*a*) at first there may be symptoms of a *unilateral cord lesion*, which are followed by

(*b*) symptoms of a *transverse lesion of the cord*.

Of the "*root symptoms*" the most important is pain. At first the pain is paroxysmal and of a neuralgic character, shooting along the course of the affected spinal nerve (around the trunk or in the limbs); or it may be boring, burning, or cutting in character. Later the pain may be dull and constant with exacerbations.

At first the pain may be unilateral, but it soon becomes bilateral. There is a sense of constriction at the part affected and often the pain is exceedingly severe, but it is not increased by bending the back. There is sometimes hyperæsthesia in the distribution of the nerve root affected, and this is followed by numbness, tingling and anæsthesia.

If the roots of the cervical or lumbar nerves are implicated, there is pain in the arms or legs; this may be followed by rigidity of muscles in the limbs affected; and later by paresis, paralysis, muscular atrophy and anæsthesia in the parts to which the nerve roots are supplied.

There is seldom any pain on percussion of the vertebral spines, though there may be pain in the back a little below the level of the growth, and also localised rigidity of the spinal muscles.

Schultze has shown that in meningeal tumour pain is occasionally absent or very slight at the early period and even later.

Symptoms of *compression of the spinal cord* finally develop, though it may not be for months after the root symptoms have become severe. The cord symptoms are those of a paraplegia which develops gradually. In rare cases when the growth causes a true myelitis at the seat of compression, the paraplegia may develop subacutely.

Sometimes there are symptoms of a unilateral lesion at first (Brown-Séquard's paralysis). When the lesion is above the lumbar region one leg is paralysed first, whilst on the opposite side there is anæsthesia below the lesion. When the lesion is in the lumbar or sacral region the unilateral motor and sensory symptoms are on the same side.

The unilateral symptoms are usually not well marked, and soon both sides are affected and paraplegia is produced. When the tumour is above the upper lumbar region the legs usually become spastic, there is rigidity to passive movements, the knee-jerks are increased, ankle-clonus develops, and the plantar reflex is of the extensor type. The bladder and rectum become paralysed and bed-sores may develop. Often the legs become markedly contracted and flexed at the hips and knees. Anæsthesia develops in the legs and trunk, the upper level of which will vary according to the level of the lesion. The knee-jerks and ankle-clonus are usually absent when the lesion is in the lumbar region or cauda equina; also if the lesion should completely destroy every nerve fibre and cell in the transverse area of the cord at the seat of growth the reflexes are usually absent below the lesion, as in other cases of transverse lesion of the cord.

A growth on one side in the cervical region may cause anæsthesia and atropic paralysis of one arm with anæsthesia of the opposite side of the body below the lesion; but soon all four limbs become anæsthetic. The reflexes are lost at the level of the lesion, and below the lesion the condition of the reflexes corresponds to that found in other transverse lesions (*see* myelitis). When the lumbar region is involved by the growth the muscles of the legs undergo great wasting and present the reaction of degeneration.

The condition of the bladder and rectum and other symptoms correspond to those met with in transverse lesion of the cord from various causes.

The author has observed cases of spinal tumour in which there was paralysis of both legs, but for some time the knee-jerks were neither

diminished nor increased, and ankle-clonus was absent. According to A. Turner there is less rigidity of the leg and spastic condition in extra-dural than in intra-dural meningeal growths.

Schultze has shown that pain is occasionally absent or slight in meningeal tumour. When, however, there are symptoms indicating that a lesion of the spinal cord is progressive, but that its upper limit remains stationary, or at least ascends only very little, we are justified in suspecting an extra-medullary tumour; and the case should be treated as such even though there may have been no irritative symptoms during the whole course of the disease (Schultze and Stursberg). In two of such cases, under the care of Schultze, an operation was performed and a meningeal growth successfully removed.

II. *Intra-medullary tumours* (growths beginning within the cord substance).

If the growth begins in the cord, near the posterior horn of grey matter radiating pain (root symptoms) may be present; but radiating pains (root pains) are usually, but not always, absent or slight in intra-medullary tumours. The symptoms are those of a transverse lesion of gradual onset—gradually progressing paraplegia and anæsthesia. The paralysis is often bilateral from the first. Sometimes, however, there are symptoms of a unilateral spinal lesion (Brown-Séquard) paralysis at first, but very soon the symptoms become bilateral.

Extensive muscular atrophy is more marked in tumours within the cord than in meningeal growths (except when the lesion is in the region of the cauda equina); the reaction of degeneration may be obtained in the paralysed muscles. Extensive bed-sores are more likely to develop when the growth is within the cord, and often there is analgesia and thermo-anæsthesia before tactile anæsthesia.

Tumours involving the posterior columns of the cord may give rise to ataxia and loss of knee-jerks.

Tumour growths involving the *cauda equina* cause pain, often very severe, in the region of the sacrum, bladder and rectum, and along the course of distribution of the sciatic nerves—bilateral sciatica. The muscles of the legs supplied by the sacral plexus become paralysed, and atrophied and may present the reaction of degeneration. The bladder and rectum become paralysed. Anæsthesia develops in the region supplied by the sciatic plexus (*see* p. 56), or in some limited portion of this area, as the gluteal region, or in this part and the back of the thighs according to the extent of the lesion. The plantar reflexes and the tendo Achillis reflexes are usually absent, but the knee-jerks are generally present (*see* p. 182).

Meningeal and intra-medullary tumours do not always follow respectively the courses here described. Intra-medullary tumours occasionally cause distinct root symptoms; also, as already mentioned, sometimes root symptoms are absent or slight in meningeal tumours.

Spinal tumours terminate in the same manner as transverse myelitis,

through cystitis and secondary kidney affections, through septic infection from a bed-sore, through ásphyxia from respiratory paralysis in lesion of the upper cervical region, or through other complications. The prognosis is always very unfavourable. Partial recovery may occur in syphilitic tumours, but in other forms the termination is fatal unless the tumour be removed surgically. Death usually occurs in one to three years.

Diagnosis.—It is necessary (1) to diagnose tumour from other lesions; (2) to decide whether the growth is intra-medullary or meningeal; (3) to localise the level of the lesion (segmental diagnosis); (4) to decide the pathological nature of the growth if possible.

The *diagnosis* is often difficult. An important point is the history of the steadily progressive nature of the symptoms. A paresis of the legs, of gradual development, which day by day becomes more marked until there is complete paralysis, and is associated with steadily increasing anæsthesia and loss of power over the bladder and rectum, should cause the diagnosis of tumour to be carefully considered, when there is no evidence of vertebral caries or of spinal injury.

Schultze has drawn attention to the fact that usually in spinal tumour, in spite of the advance of the symptoms, they do not extend upwards, or at least only to a very slight extent; since the growth does not usually extend much upwards in the cord. The symptoms of spinal tumour are those of a steadily progressing transverse lesion without indications of extension of the lesion upwards, or at least only to a slight extent. Schultze thinks this feature is of great diagnostic importance.

In *meningeal* growths pain and symptoms of nerve-root irritation, limited to one or two segments of the cord, and followed in course of time by steadily increasing paralysis, are of great diagnostic importance, (signs of caries or injury being absent). The root symptoms may be present for a long time before cord symptoms develop.

Allen Starr points out that the pain is often localised in the peripheral termination of the nerve root compressed, and is hence frequently referred to the epigastrium, abdomen, or limbs, rather than to the spine. At first the pain may be unilateral, but it soon becomes lateral, and it is rare to meet with unilateral pain when paralysis is present. There is no pain in the spine as a rule, but if the vertebræ be eroded by the growth there may be pain for one or two inches below the tumour.

Starr thinks that the order of development of symptoms is of importance. This he states to be as follows: (1) pain (root symptoms); (2) increase of the reflex excitability; (3) progressive loss of power—paresis, paraplegia; (4) anæsthesia; (5) loss of reflex activity. Brown-Séquard's paralysis occurs in some cases of meningeal tumours, but more frequently there is progressive paraplegia.

The diagnosis is most difficult in *intra-medullary* tumours; but here the slow development of the paralysis, anæsthesia, and other symptoms of a transverse cord lesion is the most important point. Pain

and root symptoms are usually, but not always, absent. Rapidly progressive muscular atrophy is more common than in meningeal tumours. Anæsthesia to pain and temperature whilst tactile sensation is preserved would be in favour of the intra-medullary seat of the tumour.

When spinal tumour is suspected, it is important to examine carefully for signs of tumour growth elsewhere ; and for evidence of tubercular disease or syphilis.

From myelitis the diagnosis is usually easy. In most cases of myelitis the onset of paralysis is much more rapid than in spinal tumour. In spinal tumour of the intra-medullary form the resemblance of the symptoms to those of myelitis is greatest ; but the history is usually diagnostic. In myelitis the onset of the paralysis is usually rapid, reaching its height in a few hours or days in acute cases, whilst in tumour the paralysis and anæsthesia develop very slowly, and day by day become more marked. Bed-sores develop late in spinal tumour, but not infrequently they occur early in myelitis. In the meningeal tumour the root pains and root symptoms developing before the onset of the paralysis are characteristic : these root pains and root symptoms do not occur in myelitis ; also the gradual development of the paralysis in most meningeal tumours distinguishes the cases from acute myelitis.

In spinal caries root pains are not so severe, the paralysis is nearly always bilateral from the first, or becomes bilateral exceedingly rapidly. In caries there are usually signs of bone disease,—an irregular or prominent vertebral spine, with pain and tenderness at the seat of the prominence. The pain in caries is decidedly increased by movement, but not in meningeal tumour as a rule ; and when it is increased by movement in the latter disease, the increase is slight. Tubercular disease in other parts of the body and psoas abscess are in favour of caries : tumour growth in other parts of the body, in favour of spinal tumour. Examination with the X rays may give evidence of caries.

In cervical pachymeningitis the pain is bilateral from the first and has a much greater vertical extent. The long course and the improvement are in favour of pachymeningitis.

From syringomyelia, spinal tumour is distinguished by the root symptoms (when present) and by the more rapid course, in the latter affection.

The symptoms in tumour of the cauda equina may be mistaken for sciatica. But the pain in the former is bilateral, and bilateral sciatica should always arouse suspicions of tumour of the cauda equina and lead to examination for other symptoms such as bilateral paresis and anæsthesia in the distribution of the sciatic plexus. Such bilateral symptoms are not present in sciatica.

The diagnosis between spinal meningeal tumour and growths in the vertebræ may be impossible at first. Tumour or deformity of the spine posteriorly would indicate vertebral growth, but these signs are often absent at first. Pain on movement is greater when the growth com-

meninges in the vertebra, and there is also pain on spinal pressure or percussion at the seat of the vertebral growth. There is usually pain in the spine as well as in the nerve distribution in vertebral growth. Examination with the X rays may be of service and may demonstrate a growth in connexion with the vertebra.

The diagnosis between meningeal (extra-medullary) and intra-medullary tumour is often very difficult. In favour of extra-medullary or meningeal tumour would be root pain or other root symptoms for months before other symptoms develop. These root symptoms are often unilateral at first. Then cord symptoms follow; they also may be unilateral at first, but soon they become bilateral. The symptoms of intra-medullary tumour are those of a transverse spinal lesion of gradual onset, and root symptoms are usually absent or slight. Dissociation anæsthesia—loss of sensation to pain and temperature whilst tactile sensation is preserved, would be in favour of an intra-medullary tumour. According to Schultze intra-medullary tumours are rare in the dorsal region of the cord. It has been already mentioned that root pains are occasionally absent in meningeal tumours.

It is desirable to decide the nature of the growth, if possible, and this is sometimes an indication as to the probable seat of the tumour. It is important to examine for signs of tumour growth in other parts of the body, and to examine for evidences of tubercular disease or syphilitic infection.

In cases of a solitary tubercular mass in the cord, there are signs of tubercular disease in other parts of the body, but no indications of vertebral caries. The symptoms consist chiefly of unilateral paralysis which rapidly become bilateral: at first there is anæsthesia for pain and temperature, followed later by total anæsthesia. Motor and sensory irritative symptoms are not prominent, and when present precede the paralysis for a short time only (Rystedt).

Metastic tumours are usually extra-medullary. Gliomata and most tubercular tumours are intra-medullary. Sarcoma may be either.

In two cases of meningeal sarcoma, recorded by Rindfleisch and Dufour, the cerebro-spinal fluid, obtained by lumbar puncture, contained cells similar to those of the tumour growth.

After concluding that the case is one of tumour it is necessary, especially if operative treatment seems desirable, to localise the lesion exactly as regards the segment of the cord affected. For this purpose the upper limit of the sensory symptoms should be exactly determined. (The segments of the cord corresponding to various levels of anæsthesia have been already described, *see* p. 97.) It is important to remember that the tumour has usually been found (post mortem or at the operation) higher than the level at which it has been localised.

Horsley has employed the hypodermic injection of pilocarpine ($\frac{1}{10}$ grain) for the localisation of the lesion. Owing to the paralysis of the vaso-motor nerves, from pressure of the tumour, sweating will not occur

in the skin area supplied by these nerves, whilst above there is profuse sweating.

The table on p. 104 shows the spinous processes corresponding to various segments of the cord.

Treatment of Spinal Tumours.

By **medical treatment** we can only hope to obtain improvement in two forms (using the term in its clinical sense), viz., tubercular and syphilitic tumours.

When we have indications of tubercular disease in other parts of the body, and when it is probable that the spinal growth is tubercular, then the general and medical treatment for tuberculosis is desirable.

When it has been decided that the spinal symptoms are due to a gumma, antisyphilitic treatment should be carried out thoroughly (*see* p. 400), and satisfactory results may follow. But it is seldom that a spinal tumour is gummatous. If we are uncertain as to the nature of the spinal tumour and if there are evidences of past syphilis, or if there is a history of exposure to the risks of syphilitic infection, it is advisable to give antisyphilitic treatment at once. But if there is no evidence of syphilis or if syphilitic infection is improbable and the diagnosis of growth clear and its localisation definite, it is not advisable to delay operation simply for the sake of giving antisyphilitic treatment a trial, seeing that gumma of the spinal meninges is so rare. The delay of an operation for several weeks, simply to give antisyphilitic treatment a trial, because syphilis though improbable cannot be excluded, may allow the compression of the cord and consequent degeneration to become too great to be relieved by operation.

When, therefore, the diagnosis of spinal tumour is clear, and the tubercular or gummatous nature of the growth improbable, the question of operation requires to be carefully considered.

If the patient decides not to be operated upon, or if the case is unsuitable for operation from the situation of the growth, or other causes, then the general medical treatment should be the same as that of myelitis.

It is important to prevent the formation of bed-sores by the use of a water-bed, to prevent cystitis by the use of the catheter, and to wash out the bladder with mild antiseptic solutions when cystitis has developed (*see* p. 138). For the pain, injections of morphia or cocaine may be necessary. In some cases pain has been relieved by placing the patient in the sitting or upright position in bed. For the troublesome spasmodic contractions of the legs, bromides or hyoscine hydrobromate may be of service.

Operative Treatment.—The celebrated case, recorded by Sir William Gowers and Sir V. Horsley in 1888, first showed that by operative treatment, it was possible to successfully remove a spinal tumour compressing the cord. The patient suffered from "complete paraplegia, motor and sensory, of slow development, accompanied by attacks of agonising spasm." A diagnosis of tumour was made by Sir William Gowers,

and operation advised. The tumour was removed by Sir V. Horsley; in course of time the paraplegia disappeared, and the result has been the recovery of the patient. The tumour was an intra-dural myxo-fibroma, about the size of a split almond, which had so compressed the cord in the upper dorsal region as to reduce its thickness to about one-half.

During the eighteen years which have elapsed since this case was recorded a considerable number of spinal tumours have been removed surgically. In only a few of the cases has permanent recovery been obtained; but the successful cases are steadily, though very slowly, increasing; and it appears probable that the operative treatment of spinal tumours will give better results in the future than that of brain tumours. This was clearly indicated by the results of operative treatment of tumours of the brain and spinal cord, recorded by various physicians and surgeons, at the congress of German scientists and physicians, 1906 (*see Neurol. Centralblatt,* 1906, p. 969). Prof. Schultze reported 4 complete recoveries in 11 operations for spinal meningeal tumour and Prof. Oppenheim 5 favourable results in 11 operations. Though the results are often unsatisfactory, it is to be remembered that without operative treatment the disease always terminates fatally.

As already described spinal tumours may arise in four situations: (1) In the vertebræ; (2) outside the dura-mater—between the outer surface of the dura and the bone of the vertebral canal—extra-dural meningeal tumour; (3) within the dura-mater, between the cord and inner side of the dura—intra-dural meningeal tumours; (4) within the cord substance—intra-medullary.

The nature of the meningeal growth and its situation in 110 cases of meningeal tumour is shown in the following table.

	Extra-dural.	Intra-dural.	Total.
Sarcoma	18	10	28
Hydatid cyst	15	1	16
Myxoma	1	12	13
Tubercle	4	4	8
Fibroma	1	11	12
Fibro-myxoma	0	1	1
Fibro-sarcoma	3	1	4
Fibro-myxo-sarcoma	0	1	1
"Connective tissue" tumour	2	0	2
Psammoma	0	8	8
Lipoma	5	0	5
Fibro-chondro-lipoma	1	0	1
Endothelioma	2	0	2
Myeloma	1	0	1
Exostosis	1	0	1
Lymphangioma	1	0	1
Chondro-sarcoma	1	0	1
Carcinoma	2	0	2
Syphilitic gumma	0	2	2
Cysticercus	0	1	1
	58	52	110

It is only in meningeal growths, groups 2 and 3, that operation is likely to be of service. In non-malignant growths the likelihood of permanently successful results is much greater than in malignant tumours.

In 70 cases of operation for spinal meningeal tumour the nature of the growth and its situation were as follows :—

	Extra-dural.	Intra-dural.	Total.
Sarcoma	14	4	18
Hydatid cyst	17	0	17
Psammoma	0	5	5
Fibroma	1	5	6
Myxoma	1	0	1
Fibro-myxoma	0	2	2
Fibro-sarcoma	3	5	8
Fibroma or Fibro-sarcoma	0	1	1
Fibro-myxo-sarcoma	0	1	1
"Connective tissue" tumour	2	0	2
Endothelioma	2	1	3
Myeloma	1	0	1
Exostosis	1	0	1
Lymphangioma	1	0	1
Lipoma	1	0	1
Chondro-sarcoma	1	0	1
Cancer	1	0	1
	46	24	70

It is interesting to note that this table does not contain tubercular or gummatous tumours.

The prospects of successful surgical treatment are best in hydatid cysts and other rare forms of cysts : in other cases the prospects of cure are much less. The results are usually unsuccessful because :—

(1) Frequently the growth commences in the bone (vertebræ).

(2) The growth is most frequently malignant and recurs after operation.

(3) The growth is often too extensive to be removed.

(4) If removed, the operation is often done too late and the cord has been too much compressed for recovery to occur. The diagnosis is often made late ; or the patient long hesitates to submit to the operation. Thus valuable time is lost.

At first, in meningeal growths, the spinal symptoms are due to local œdema of the cord, and if the growth could be removed at this period the nerve structures would recover. Later the nerve fibres and cells degenerate, and recovery does not occur even when the growth is removed. This last difficulty (4) can be overcome by early diagnosis and early operation, but the other difficulties cannot be overcome.

Hence operative treatment is only likely to be successful in a small number of cases ; but it is the only treatment which can cure the patient, except in the case of tubercular and gummatous tumours.

Collins estimates that 50 per cent. of spinal meningeal tumours are suitable for operation.

When the diagnosis of spinal meningeal new growth (excluding tubercular and gummatous tumours) has been made, an exploratory operation is desirable, in most cases, as early as possible. If the growth is found to be unsuitable for removal, the operation should be discontinued.

For the successful operative treatment of spinal tumour not only is a correct differential diagnosis necessary, but the growth must be exactly localised by a consideration of the upper limit of the motor and sensory symptoms—especially the upper limit of the pain and anæsthesia. From the evidence furnished thereby, the level of the upper limit of the lesion is diagnosed, and the vertebral spine corresponding to this segment of the cord is first removed.

It has been pointed out by many surgeons that the tumour was found higher than was anticipated from the symptoms. Hence Bruns states that "if symptoms of a sensory nature point to any one dorsal segment of the cord being pressed upon by a tumour, the operation should expose the dorsal segment one or even two levels higher." Starr points out that the posterior nerve roots run upward for a considerable distance on entering the spinal canal, before entering the spinal cord, and also that they turn up within the cord before ending in the posterior horns. Hence pain may be caused by pressure at a level considerably higher than the level of the pain. In a case reported by Starr, the level of the pain was about 8 inches lower than the level of the tumour. Hence he concludes that in operation for spinal tumour in the dorsal region a level of the cord should be exposed at least 4 inches higher than the upper limit of the anæsthesia in the back, and that in case no tumour is then found, the wound should be enlarged upwards.

Allen Starr recommends that the dura should be exposed over a region at least 2 inches long, and if palpation does not reveal the tumour, it is well to lay bare another inch before opening it. It is better to go higher than lower. The dura should pulsate, in the normal condition, but does not do so near a tumour, and especially below it. Starr thinks it is well to open the dura in all cases, as sometimes a second growth is found on its inner side. He considers that it is important to keep the patient in the prone position, or on the side, as long as possible after the operation, to prevent undue drainage of cerebrospinal fluid.

Death after the operation sometimes occurs through shock, hæmorrhage, or septic meningitis. But the dangers of the operation are not so great as in cerebral tumour. The greatest danger is septic infection and septic meningitis, especially when the dura mater has been opened. Respiratory failure is a serious complication in tumours in the cervical region of the cord.

When the tumour cannot be removed division of the posterior nerve roots, both above and at the region of the growth, is of service for the relief of pain.

A careful analysis of the results of surgical treatment of spinal tumour has been made by R. H. Harte. In 92 cases (collected from literature)

he found that 29 are recorded as cured, 17 as improved, 3 as not improved. 49 cases recovered from the operation : 43 died. Of course, many of the patients who recovered from the operation died at a later period : and in many cases reported as cured, it is probable that there would be a recurrence of the growth later.

In cases of tumour of the vertebræ compressing the cord operative treatment will not give permanent relief except in very rare non-malignant forms of growth. In the ordinary form of sarcoma of the spine operative interference is generally useless, but Sir V. Horsley points out that it may be undertaken, if necessary, to relieve extreme pain, since this is produced purely mechanically. Horsley states, that cases of primary sarcoma considered inoperable may with advantage be treated with the injection of Coley's fluid, and local necrosis thus induced with consequent relief of pressure.

It is impossible, in the present state of surgery, to remove intramedullary tumours without too great injury to the rest of the cord. When there is difficulty in deciding whether a spinal growth is intramedullary or meningeal, it is advisable to recommend an exploratory operation, if there are any symptoms in favour of the latter, since in several doubtful cases, recently recorded, operation has revealed a meningeal growth which has been successfully removed.

I have collected from medical literature 51 cases of *meningeal* spinal tumours (including cysts) in which operation has been *successful* and has been followed by recovery from the spinal symptoms, completely or to a great extent. Six cases of vertebral tumour are also included. There are many other cases on record in which the growth was removed successfully, but as recovery from the symptoms did not occur they are not included in the following list.

OPERATIVE TREATMENT OF SPINAL TUMOURS (MENINGEAL). SUCCESSFUL CASES.

Nature of Tumour.	Extra-dural.	Intra-dural.	Total.
Hydatid cysts	10	0	10
Fibroma	1	4	5
Fibro-myxoma	0	2	2
Myxoma	1	0	1
Fibro-sarcoma	3	4	7
Fibro-myxo-sarcoma	0	1	1
Fibroma or Fibro-sarcoma	0	1	1
"Connective tissue" tumour	2	0	2
Chondro-sarcoma*	2	0	2
Psammoma	1	3	4
Exostosis*	1	0	1
Sarcoma	5	2	7
Endothelioma	1	1	2
Lymphangioma	1	0	1
Cyst (not parasytic)	1	1	2
Myeloma*	2	0	2
Nature not stated	0	1	1
* indicates vertebral tumour	31	20	51

As regards the nature of the growth, permanent success may be expected in cases of fibroma, myxoma, fibro-myxoma, psammoma, lymphangioma, exostosis, and perhaps also in fibro-myxo-sarcoma and fibro-sarcoma. Also hydatid cysts (as shown by the table) may be expected to be removed successfully, when the diagnosis is made early.

Prof. Schultze of Bonn has had a fortunate experience. In 13 of his cases of meningeal spinal tumour an operation has been performed, and in 6 complete recovery, and in 1 permanent improvement, have followed (i.e. 50 per cent.).

In 8 of these cases, in which there was a non-malignant tumour in the dorsal region, operation was followed by complete recovery in 6 and permanent improvement in 1.

It is interesting to note that in two of Schultze's cases, in which a meningeal tumour was removed and recovery followed the operation, there had been no root pains or other kind of pain; also in several other cases a meningeal growth was found and removed successfully, when the diagnosis had been doubtful.

Oppenheim has recorded 5 out of 11 cases of this operation in which a successful result was obtained.

The following tables show the position of the growth, its pathological nature, and the results of the operation, in 51 successful cases.

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Extra- and Intra-dural Meningeal Spinal Tumours Successfully Removed by Operation.

Regions.	Pathological Nature.	Results.	Authors.	Journals.
1. Intra-dural, 4 & 5 dorsal segments	Fibromyxoma .	Recovery complete	Gowers & Horsley	*Med. Chir. Trans.*, 1888, p. 407.
2. Extra-dural, under the spines of 5th to 7th dorsal vertebræ	"Connective tissue mass"	Recovery. Reported 5 years after operation	McEwen	*Lancet*, Aug. 11, 1888.
3. Extra-dural, dorsal	"Connective tissue" tumour	Recovery. Reported 4 years later	McEwen	" " "
4. Extra-dural, cauda equina	Lymphangioma .	Recovery . . .	Laquer & Rehn	*Archiv. f. klin. Chir.*, Bd. 42, p. 812.
5. Extra-dural .	Exostosis . . .	Recovery . . .	Hahn	Mentioned *Neurologisches Centralblatt*, 1902, p. 621.

Regions.	Pathological Nature.	Results.	Authors.	Journals.
6. Extra-dural, dorsal	Hydatid cyst .	"Terminating favourably"	Horsley	*Clinical Journal*, Vol. IX., 1896–97, p. 177.
7. Extra-dural, dorsal	Hydatid cyst .	Recovery . . .	Lloyd	*American Med. and Surg. Bull.*, New York, 1896, X., p. 659, quoted by Putnam & Warren.
8. Extra-dural	Hydatid cyst	Recovery	Szekeres	*Pester Med. Chir. Presse*, 1894, p. 43. Quoted by Putnam & Warren.
9. Extra-dural & vertebral	Hydatid cyst .	Recovery . . .	Hahn	*Centralblatt f. Chir.* (Society Report), 1902, No. 14, p. 398.
10. Extra-dural, dorsal	Hydatid cysts .	Recovery: almost complete	Tytler & Williamson	*British Medical Journal*, Feb. 7, 1903.
11. Extra-dural, adherent to 5th & 6th dorsal laminæ	"Fusiform" spindle cells, sarcoma	Recovery, doing work 7 months after operation	Davies-Colley	*Trans. Clin. Soc.*, Lond., 1892, p. 163.
12. Intra-dural, dorsal	Psammoma . .	Marked improvement (12 months later could walk without aid of stick)	Lichtheim & Mikulicz	*Deutsche Med. Woch.*, 1891, p. 1386.
13. Intra-dural, dorsal	Psammoma . .	Recovery almost complete after 10 months	F. Krause (also reported by Boettiger)	*Berliner klin. Woch.*, 1901, Nos. 20–22.
14. Intra-dural, dorsal	Fibroma . . .	Almost complete recovery after 2 months	Oppenheim & Jolly	*Abstract Neurologisches Centralblatt*, 1902, p. 619.
15. Intra-dural, dorsal	Fibroma . . .	Recovery . . .	Eskridge & Freeman	*Philadelphia Med. Journ.*, Dec. 10, 1898. Quoted by Putnam & Warren.
16. Intra-dural, dorsal	Fibroma . . .	Complete recovery: recorded at end of 7 years	Putnam & Warren	*American Journal of the Med. Sciences*, Oct., 1899, and *Journal of Nerv. and Mental Dis.*, 1902, p. 100, and *Boston Med. and Surg. Jour.*, July 20, 1905.
17. Extra-dural, involving upper cervical vertebræ	Sarcoma . . .	Able to walk 4 years later	Putnam & Elliott	*Journ. Nerv. & Ment. Dis.*, 1902, p. 100.
18. Intra-dural, lower cervical region	Sarcoma . . .	Improved at end of 1 year and 9 months. Progressing well at end of 3 years.	Abbe	*Journal Nervous and Mental Diseases*, 1902, p. 281; 1903, p. 103.
19. Extra-dural cauda equina	Fibro-sarcoma .	3 months later marked improvement; doing well 3 years later.	Sachs	*Med. Record*, Jan. 6, 1900, and *Journ. of Nerv. and Mental Diseases*, 1903.
20. Extra-dural, cauda equina	Alveolar-sarcoma	Recovery: 2 years later described as successful; doing well 1 year later	Sachs	*Med. Record*, Jan. 6, 1900, (mentioned *Journal Nerv. and Mental Diseases*, 1902 and 1903.
21. Extra-dural, invading dorsal vertebræ	Sarcoma . . .	Great improvement, disappearance of paralysis, patient able to do work at the end of 3 months	Kümmell	*Archiv. f. klin. Chir.* 1895, p. 458.

Regions.	Pathological Nature.	Results.	Authors.	Journals.
22. Extra-dural, dorsal	Fibro-sarcoma .	Can walk with a stick at end of 14 months	Schultze & Schede	Mentioned *Berliner klin. Woch.*, 1901 No. 39, p. 1006, and *Neurologisches Centralblatt*, p. 634, 1902.
23. Intra-dural, dorsal	Fibro-myxo-sarcoma	Can walk with a stick at end of 6 months	Schultze & Schede	,, ,, ,,
24. Intra-dural, dorsal	Fibro-sarcoma .	Can walk at end of 7 months	Schultze & Schede	,, ,, ,,
25. Extra-dural, cauda equina	Endothelioma .	Partial recovery (reported soon after operation)	A. Starr	*Journal of Nervous and Mental Diseases*, 1901, p. 157.
26. Extra-dural, dorsal	Chondro-sarcoma	Improved . . .	Vynaloff	Quoted by Putnam & Warren, l. c.
27. Extra-dural, dorsal, connecnected with vertebræ	Myelom . . .	Recovery and no return of symptoms at the end of 6 months. Coley's fluid also used.	J. J. Thomas & Munro	*Journal of Nervous and Mental Diseases*, p. 98, 1902.
28. Meningeal, intra-dural, 5th cervical to 1st dorsal vertebra	Fibro-sarcoma .	Marked improvement; could walk at end of 10 months after operation	Henschen & Lennander	*Upsala Läkareförenings Förhandlingar*, p. 473, 1901, (abstract *Med. Review*, 1902, p. 571.)
29. Tumour of 6th dorsal vertebra, extra-dural	Chondro-sarcoma	Marked improvement; could walk with aid of stick 95 days after operation	Israel	*Berliner klinische Woch.*, No. 22, 1903.
30. Cauda equina	? nature: sarcoma or granulation tissue	Recovery; recorded 2½ years later	Bailey & McCosh	*Journ. Nervous and Mental Diseases*, Feb. 1903, p. 99.
31. Intra-dural meningeal, lower cervical region (left side)	Fibro-sarcoma .	Recovery practically complete 3 months after operation	Cushing	*Annals of Surgery*, June 1904.
32. Intra-dural, meningeal, in region of 6th cervical lamina (on the left side)	Endothelioma .	Great improvement 7 months after the operation; patient able to walk without assistance; considerable return of power in left arm	Woolsey	*Med. News*, New York, Oct. 1, 1904.
33. Intra-dural, cauda equina, arising from 4th lumbar posterior nerve root on the right side; compression of lower end of cord	Fibro-myxoma .	Recovery almost complete; could walk well 12 months after operation	Sir V. Horsley	*Report of Meeting of Neurological Society*, London, *Brain*, 1904.
34. Extra-dural, 4th dorsal segment	Hydatid cyst .	Complete recovery; was able to walk at end of 7 months	Turner, A.	*Clinical Journal*, June 8, 1904.
35. Extra-dural, 6th dorsal segment	Myxoma . . .	Recovery; able to work at time of publication	Turner, A.	*Clinical Journal*, June 8, 1904.
36. Intra-dural, 3rd cervical segment	Round-celled sarcoma	Recovery; distinct improvement in paralytic and sensory symptoms 10 weeks after operation	Putnam, Krauss, Park	*American Journal of the Med. Sciences*, Jan., 1903.

Regions.	Pathological Nature.	Results.	Authors.	Journals.
37. Intra-dural, lumbar	Cyst (not parasitic)	Cured; reported 9 months after operation	Spiller, Musser & Martin	*University of Pennsylvania Med. Bull.* Mar. and April, 1903.
38. Extra-dural, dorsal	Cyst (not parasitic)	Cured; reported 8 months after operation	A. Schmidt	*Deutsche Zeitschrift f. Nervenheilkunde*, 1904, p. 318, Bd. 26.
39. Extra-dural, dorsal	Hydatid cyst .	Improved, 2 months after operation. Ultimate result (?)	A. G Owen, case of Sir W. Gowers and Sir V. Horsley	*Inter-Colonial Medical Journ.*, Dec. 20, 1905.
40. Extra-dural and vertebral, lumbar	Hydatid cyst .	Improved, 6 months after operation	A. G. Owen, case of Sir. V. Horsley	Ditto
41. Extra-dural, lumbar	Hydatid cyst .	Improved, 4 months after operation	A. G. Owen, Ormerod, Sir V. Horsley	Ditto
42. Extra-dural, dorsal	Hydatid cyst .	Recovery 4 years after operation. Could walk well, but legs slightly spastic	A. G. Owen, Bastian, Sir V. Horsley	Ditto
43. Intra-dural, cervical	Fibro-sarcoma .	Almost complete recovery 6 months after operation	Brodnitz & Auerbach	*Neurologisches Centralblatt*, 1905, 619 (abstract).
44. Intra-dural, dorsal	Fibroma . . .	"Practically complete recovery"; last report 7 years after operation	J. C. Warren	*American Medicine*, Aug. 26, 1905.
45. Intra-dural, dorsal	Psammoma . .	"Practically well" 6 months after operation	J C. Warren	Ditto
46. Intra-dural, cervical	Fibroma or fibro-sarcoma	Eight months after operation could walk quite well and do housework, but left arm remained paralysed.	Oppenheim and Borchardt	*Berliner klin. Woch.*, June 25, 1906.
47. Intra-dural, dorsal	Not stated . .	Great improvement, able to walk at end of 2 months, but ataxic	Oppenheim & Borchardt	Ditto
48. Extra-dural, cervical	Fibro-sarcoma .	Paralysis of bladder and rectum, and anæsthesia soon disappeared; could walk alone 12 months after operation	H. C. Baldwin	*Boston Med. and Surg. Journal* May, 31, 1906, and Dec. 6, 1906, p. 677.
49. Extra-dural, growth from laminæ of vertebræ, lower cervical region	Myeloma . .	Relief of pain, rapid improvement of motor and sensory symptoms. Growths appeared later in other parts of body	Walton & Paul	Ditto, July 2, 1905. Vol. ii.
50. Extra-medullary (attached to dura ? extra-dural) dorsal	Psammoma . .	Recovery from paralysis; able to walk without the aid of a stick	(Schultze) Stursberg	*Deutsche Zeitschrift. Nervenheilkunde*, Bd. 32, 1907.
51. Extra-dural, dorsal	Fibroma . . .	Complete recovery	Schultze	*Münchener med. Woch.* No. 28, 1907.

SPINAL CYSTS—HYDATID, CYSTICERCI, AND SIMPLE CYSTS

Other Rare Forms of Compression Myelitis.

The simple or non-parasitic cysts are extremely rare. They are meningeal and contain clear serous fluid. (Two cases removed successfully are mentioned on p. 178.)

The *parasitic cysts* are cysticerci cellulosæ and hydatid cysts. The latter are more common than the former. Cysticerci are usually meningeal and intra-dural, but very rarely intra-medullary. They cause compression myelitis. Usually similar cysts are found in other parts of the body.

Spinal Hydatid Cysts.

Affection of the spinal cord owing to the pressure of hydatid cysts is exceedingly rare. The cysts causing the spinal compression are usually situated *external to the spinal dura mater* (20 out of 24 cases collected by Colman), and generally they develop in the loose adipose tissue between the dura mater and the bone of the vertebral arches. In very rare cases the cysts have been found within the dura mater. In many of the cases recorded the cysts have first developed in the subpleural or subperitoneal connective tissue, or in the muscles of the back ; afterwards they have extended through the intervertebral foramina into the vertebral canal, and have then spread in the longitudinal direction in the adipose tissue surrounding the spinal dura mater. (This had occurred in a case reported by Dr. Tytler and the writer.) In some cases the bone of the vertebræ has been affected primarily or secondarily by the cyst : the bone has been eroded, and in rare cases spinal curvature has been produced by collapse of the affected vertebræ. In other rare cases an external fluctuating spinal swelling has been caused by the cyst.

Maguire has pointed out that in by far the majority of cases the cysts are situated at the posterior surface of the cord.

The cysts are always unilocular (Schmaus) : their size varies from a pea to a walnut, and they are often numerous.

The region of the cord compressed in 25 cases collected by Colman was as follows : Cervical, 2 ; dorsal, 10 ; lumbar, 6 ; cauda equina, 5 ; dorsal, lumbar, and cauda equina, 2. Colman also gives the site of origin of the hydatid cysts in 21 cases as follows : Cancellous tissue of the vertebræ, 7 ; extra-dural areolar tissue, 4 ; arachnoid tissue, 2 ; muscles of the back, 4 ; lung or pleural cavity, 2 ; retroperitoneal tissue, 2.

Colman's analysis of cases recorded gives the following results, as regards the presence of cysts in other parts : Not stated, 19 cases ; no other cysts, 8 cases ; cyst in dorsal muscles, 2 ; in areolar tissue of pelvis, 1 ; in liver, 2 ; in back muscles, iliac fossa and prevesical areolar tissue, 1.

The spinal hydatid cysts cause " compression myelitis " in the same

way as tumour growths in the meninges or vertebræ. In some cases there is mechanical compression of the cord. But in others the compression is not direct, and the cyst simply causes obstruction to the blood and lymph circulation by diminishing the space in the vertebral canal. In both ways œdema of the cord is produced and later softening and degeneration occur.

Symptoms.—The cysts produce symptoms of compression myelitis, resembling those of meningeal tumour (*see* p. 164). In most of the cases recorded, symptoms of compression of the spinal nerve roots, consisting chiefly of radiating pains, have occurred first, and later paraplegia, anæsthesia, and bladder and rectal symptoms have developed. The onset of the paraplegia has usually been gradual.

Diagnosis.—Symptoms of compression myelitis, simulating those of meningeal tumour, can be attributed to the pressure of hydatid cysts when examination reveals the presence of such cysts in some other part of the body. In some cases a small cyst has been felt in the back, close to the spine, and when this has been punctured, the fluid has had the characters of hydatid fluid. It has been colourless or slightly opalescent (but devoid of yellow tint), and on microscopical examination hydatid scolices or hooklets have been found (*see* case mentioned at the end of this description).

Treatment.—The only treatment of any service is early laminectomy and removal of the cysts. In the cases in which this operation has been performed, usually the compression has been of long standing, and the case has terminated fatally, although the cysts may have been successfully removed. But early diagnosis and operation would have saved many of the cases recorded. In the table on p. 175, 10 successful operations for spinal hydatid cysts are mentioned.

Pathological anatomy has revealed three facts of great importance with respect to the favourable prospects of operation. (1) The cysts causing the compression myelitis are usually external to the dura mater (20 out of 24 cases) and can therefore be removed without opening the dura. (2) The cysts are usually on the posterior surface of the dura mater, and are therefore most easily exposed by laminectomy. (3) At first the cause of the paralysis is localised œdema of the cord; at a later period degeneration of nervous structures and softening occurs. If the cysts can be removed early, when the paralysis is simply due to œdema of the cord, recovery may be expected.

In a case which was under the care of Dr. P. Tytler and myself, at the Ancoats Hospital, the symptoms were: Complete paralysis of both legs, anæsthesia of the legs and abdomen and lower part of thorax up to the 5th rib, paralysis of bladder and rectum. The symptoms commenced with severe pain in the back (mid-dorsal region) which continued three weeks; then paralysis of the legs gradually developed, and in a week's time the patient was unable to walk. The symptoms suggested a compression myelitis, rather than acute transverse myelitis, and corresponded to

those of a meningeal tumour. There was a small swelling in the back, just under the skin, and to the left of the 3rd and 4th dorsal vertebral spines. The diameter of the swelling was about 1 inch : it was punctured and the fluid was found to contain hydatid hooklets. Dr. Tyler operated on the case and removed 14 hydatid cysts (which were in the vertebral canal, external to the dura mater, in the dorsal region). Improvement followed rapidly, the anæsthesia and paralysis of bladder and rectum soon disappeared, the patient gradually regained power of movement in the legs, and she was able to walk with the aid of a stick at the end of twelve months. Further improvement has steadily continued. When seen six years after the operation she felt quite well. There was still some rigidity of the legs, but this was slowly diminishing and the patient could walk to the hospital with the aid of a stick. (The case is recorded in the *British Medical Journal*, February 7, 1903. Other references are given in this article and in the table on p. 175.)

Compression by Exostoses.

In rare cases exostoses connected with the vertebræ have caused compression of the cord. Pain and other symptoms of compression of nerve roots have been followed by symptoms of compression of the cord. The diagnosis can only be made when exostoses are present in other parts of the body. In a case mentioned by Hahn [1] the exostosis was removed and the patient cured. It is probable that a few of the cases could be treated successfully by operation.

Compression of the Cord by Aneurism.

In rare cases an aneurism of the aorta has eroded the dorsal or lumbar vertebræ and compressed the spinal cord. The symptoms are chiefly pain in the back, pain in the distribution of the nerve roots compressed, and afterwards symptoms of "compression myelitis" of slow or rapid onset. If the aneurism ruptures into the vertebral canal paraplegia develops suddenly, and death occurs in a very short time. The diagnosis can only be made when the symptoms of aneurism are associated with those of "compression myelitis."

A Röntgen ray photograph or the use of the X Ray screen, may give valuable indication of an aneurism in doubtful cases.

For an account of syphilitic disease of the vertebræ *see* chapter on Spinal Syphilitis, p. 388.

DISEASES OF THE CAUDA EQUINA AND CONUS TERMINALIS (OR CONUS MEDULLARIS).

Cauda Equina.—Lesion of the nerve roots which form the cauda equina may be due to the following causes : Injury, fracture and dislocation of vertebræ, wounds, hæmorrhage, bullet wounds, tumours

[1] Centralblatt f. Chirurgie, No. 14. Society Report, p. 398, 1902.

(neuroma, sarcoma, glioma, lymphangioma, fibroma, endothelioma, etc. arising from the nerve roots, the spinal membranes, or the vertebræ), syphilitic gumma or meningitis, abscess from tubercular bone disease.

The chief symptoms of lesion of the cauda equina are [1] :—

1. Paralysis of muscles of the legs, usually limited to those supplied by the sacral plexus or the lower sacral segments of the cord.

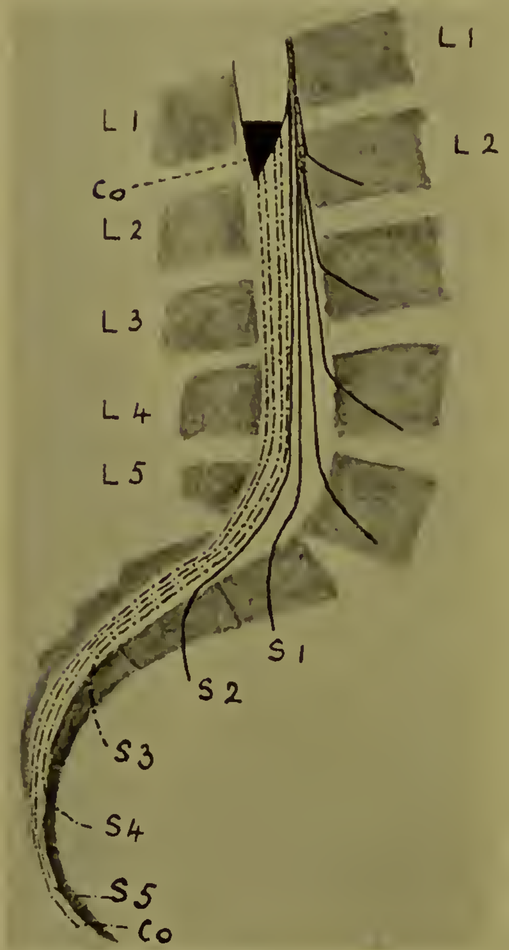

FIG. 86A.—Vertical Section of Lumbar Vertebræ and Sacrum, showing termination of Spinal Cord, and nerves of cauda equina in vertebral canal. Deep black = termination of Cord, CO = Conus terminalis. L 1 to 5 = Lumbar vertebræ. S 1 to 5 = Sacral roots. Broken lines = nerves from conus, S 3 to 5, and coccygeal nerves = CO.

2. Paralysis of bladder and rectum, with loss or diminution of sexual power.

3. Pain, often very severe, in the region of the sciatic nerves (double sciatica), and in the region of the sacrum, bladder and rectum.

4. Anæsthesia of the mucous membrane of the bladder, urethra and rectum. Anæsthesia of the scrotum and penis in the male, or of the vagina and vulva in the female. Anæsthesia of the perineum, anus and gluteal region. A saddle-shaped area of anæsthesia may be present in the gluteal region, and when the lesion is extensive, there may be a strip of anæsthesia on each side down the back of thigh, with anæsthesia on the outer side of leg and foot, i.e. in the distribution of the sacral plexus (*see* Fig. 56).

5. The plantar reflexes are lost; the tendo Achillis reflexes are usually lost; the knee-jerks and cremasteric reflexes are usually present.

6. Atrophy of paralysed muscles. Bed-sores.

7. Diminished excitability, faradic and galvanic, in the nerves of paralysed muscles, and sometims reaction of degeneration in the muscles.

When all the nerve roots of the cauda equina are affected, all the leg muscles are paralysed and undergo wasting, and the anæsthesia extends up to the groin. But usually the upper two lumbar nerves, at least, are not affected; and the anæsthesia and paralysis are less extensive, according to the level of the lesion. Often the paralysis and anæsthesia are limited to the distribution of the nerves of the sacral plexus or its inferior portion.

[1] *See* Descriptions by W. Thorburn, Raymond, Schultze and Oppenheim.

If the lesion of the cauda equina is below the exit of the 2nd sacral nerves, the symptoms are :—paralysis of the bladder and rectum and loss of sexual power (ejaculation), with a saddle-shaped area of anæsthesia in the gluteal region, and anæsthesia of the perineum, scrotum, urethra and posterior part of the thighs. At a lower level the symptoms are still more limited (*see* p. 98, spinal localisation).

Conus Terminalis.—The spinal cord terminates in a pointed or conical extremity, at its lower end, to which the name of *conus medullaris* or *conus terminalis* is given. There is no line of demarcation between the conus and the spinal cord, but Raymond has fixed the upper limit of the conus between the origin of the 2nd and 3rd sacral nerves, and this limit has been generally accepted. In front of the conus terminalis is the body of the 2nd lumbar vertebra ; behind it the spine of the 1st lumbar. A knife passed into the vertebral canal between the 1st and 2nd lumbar vertebræ would pass through the base of the conus terminalis.

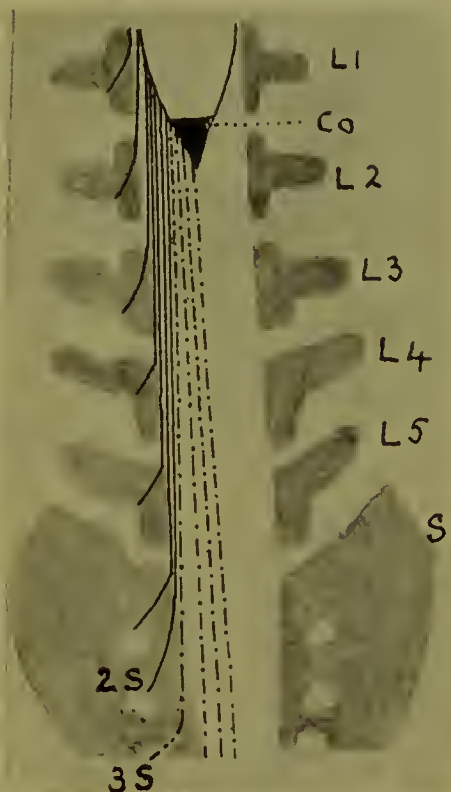

FIG. 86B. — Diagram showing position of Conus Terminalis and Cauda Equina in Vertebral Canal when opened posteriorly. Letters as in 86A.

The conus terminalis is believed to contain the centres for erection of the penis and ejaculation, the centres for discharge of the contents of the bladder and rectum, the centres for the sensory innervation of the lower part of the rectum, the anus, perineum, scrotum, penis, urethra, and bladder.

The chief pathological lesions of the conus terminalis are : Trauma (traumatic "myelitis,") tumours and gliosis, hæmatomyelia and meningeal hæmorrhage, and syphilitic affections. The symptoms of disease of the conus are :—

Paralysis of bladder and rectum.

Loss of sexual reflex action.

Anæsthesia (total or dissociate) of the skin of the ano-perineal and genital region (with exception of the testicles). Anæsthesia of urethra, bladder and lower part of rectum.

The anæsthesia in the gluteal region in the form of a saddle-shaped area—riding trousers form.

The tendo Achillis reflex is lost. The motor power of the legs may be preserved (Oppenheim).

If the lesion should extend higher, into the lumbar region just above the conus, the anæsthesia is more extensive, and atrophic paralysis of

the leg muscles is added to the symptoms already mentioned. But it is better to describe as affection of the conus, only those cases in which the lesion is below the 2nd sacral segment and the muscles of the leg are spared.

According to L. R. Müller the automatic centres for the bladder and rectum and genital organs are not in the cord, but in the ganglia of the pelvic sympathetic, and only the spinal ganglion cells innervating the external sphincters are in the conus terminalis. A number of cases of lesion of the conus and cauda equina analysed by Bálint and Benedict support this view. In all cases there was loss of voluntary control, but involuntary automatic action was observed, and not paralytic incontinence (*Deutsche Zeitschrift für Nervenheitkunde*, Bd. 30, 1905, p. 1).

Diagnosis.—In affections of the conus and cauda equina there are many symptoms which are common. The differential diagnosis is, however, of practical importance, because certain lesions of the cauda are suitable for surgical treatment, whilst surgical treatment is out of question in lesions of the conus. Though a definite diagnosis is often not possible, still there are a number of points in the symptomatology, which may enable a diagnosis of the probable seat of the lesion to be made.[1]

1. If the symptoms are not due to trauma, a sudden onset or a sudden extension of the symptoms is in favour of lesion of the conus; a gradual onset in favour of lesion of the cauda equina.

2. In traumatic cases, if the seat of the injury be the upper lumbar region of the spine, or if the spinous processes of the 12th dorsal or 1st and 2nd lumbar vertebræ be depressed or displaced, probably the lesion is in the conus. If the injury be in the inferior lumbar region or adjacent part of the sacrum, the lesion is probably in the cauda equina.

3. Pain at the level of the 12th dorsal and 1st and 2nd lumbar spines, which is produced or increased by pressure, percussion, or by the application of heat or cold, and pain which does not radiate into the legs point to lesion of the conus. Whilst pain below the level just mentioned, which is increased by pressure over the sacrum and by movement, and pain radiating into the legs point to lesion of the cauda equina.

4. Very severe pain in the distribution of the nerves of the sacral plexus (sacrum, bladder, perineum, anus and parts supplied by the sciatic nerves) is in favour of a lesion of the cauda equina; absence of pain in favour of lesion of the conus. Moderate pain may occur in either affection.

5. Pain which precedes other symptoms for a long period is in favour of lesion of the cauda equina.

6. Anæsthesia quite symmetrical in distribution, early and intense muscular atrophy, with the reaction of degeneration, and early onset of bed-sores are in favour of lesion of the conus and lower end of the cord.

7. In favour of lesion of cauda equina are asymmetrical distribution

[1] *See* writings of Thorburn, Raymond, Schultze and Oppenheim.

of the anæsthesia, progressive onset of the bladder and rectal troubles, the alternation of increase and decrease in the severity of the chief symptoms, slow onset and diffuse character of the muscular atrophy, absence of qualitative changes in the electrical reactions and absence of bed-sores.

Oppenheim thinks that marked anæsthesia is in favour of conus affection. Schultze believes that loss of sensation to pain and temperature, whilst tactile sensation is preserved, is a little in favour of lesion of the conus; but as this sensory disturbance has been sometimes observed in lesions of the cauda equina its diagnostic value is not great.

The most important symptom of the cauda equina is pain; and motor symptoms occur only after the pain has been present for a long period.

Diseases of the sciatic or sacral nerves have to be considered in the differential diagnosis; but these are mostly unilateral, whilst symptoms of cauda equina lesion are bilateral; also affections of the bladder and rectum are absent in the former.

Prognosis and Treatment.—The prognosis is more favourable in lesions of the cauda equina than in those within the cord at its lower end.

Hæmorrhage around the nerve roots of the cauda equina may in time become absorbed.

Traumatic fracture-dislocation affecting the cauda equina has been successfully operated upon by W. Thorburn, of Manchester, and others,

Tumour growths of the cauda equina have been, in five instances at least, successfully removed; the nature of the growths, being lymphangioma (Laquer and Renn), two cases of sarcoma (Sachs), endothelioma (Starr), fibromyxoma (Horsley) (*see* p. 175).

Operative treatment for abscess associated with bone disease has also been successful. Syphilitic lesion may improve under antisyphilitic treatment. In Bolton's case, and in another reported by Engelmann, the removal of a bullet from the sacral canal was followed by improvement.

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SPINAL HÆMORRHAGE—HÆMATOMYELIA

Primary hæmorrhage into the spinal cord is exceedingly rare. Many cases diagnosed clinically as primary spinal hæmorrhage have been shown to be due to myelitis or other lesion, or to myelitis with secondary hæmorrhage.

The rarity of primary spinal haemorrhage, in comparison with cerebral haemorrhage, is due to the following causes : (1) The spinal arteries are long, narrow, and tortuous, and hence they are not subject to the high pressure which is such an important cause of the degeneration and rupture of the cerebral arteries. (2) Miliary aneurisms, the rupture of which is such a frequent cause of cerebral hæmorrhage, are almost unknown in the spinal cord. (A few cases, however, are on record.) (3) Diseases of the vessels, which predispose to hæmorrhage, are less frequent and less severe in the spinal arteries. (4) There is a firmer supporting connective tissue around the spinal vessels than around the cerebral.

Though primary spinal hæmorrhage is so rare a number of cases are on record in which the diagnosis has been verified pathologically.

Etiology.—Spinal hæmorrhage may occur at any age; but the patients are usually much younger than the sufferers from cerebral hæmorrhage, the majority being between the ages of 10 and 40. Males suffer more frequently than females, owing to the greater liability of the former to external injury and muscular over-strain. Most cases of primary spinal hæmorrhage are of traumatic origin (90 per cent. Oppenheim) ; but often there is no actual lesion of the vertebral column or meninges, and the nature of the injury has varied considerably. The injury has often been a fall from a height on to the back, on the gluteal region, or on to the feet ; in other cases the cause has been a blow on the back, or the forced bending forwards of the head, as in riding through a gateway, etc.

After a difficult labour, in which delivery has been accomplished by the use of the forceps or by artificial means, spinal or medullary hæmorrhage has been occasionally found in the new born child (Schultze).

Very violent muscular exertion or strain has been the exciting cause in rare instances. It is said that spinal hæmorrhage occurs in horses from the same cause.

Great sexual excess is reported as the cause in one case. In a few cases the disease has developed during pregnancy or after parturition, or it has followed severe vomiting during pregnancy.

Sudden changes in the atmospheric pressure have occasionally caused the disease in divers (*see* caisson disease).

Spinal hæmorrhage may occur secondary to myelitis, and spinal softening (from vascular obstruction or other cause) : also it is occasionally secondary to tumour growths, especially glioma.

I have known the symptoms of primary spinal hæmorrhage to occur in a youth directly he left the water after diving in a swimming bath : and in two other cases (youths) the symptoms followed directly after violent muscular exertion. In the case which I have recorded on p. 392, syphilitic disease of the spinal blood vessels had been followed by thrombosis and softening and extensive hæmorrhage into the grey matter (*see* Plates IV and V). Rare cases have occurred in patients who have

suffered from purpura and blood diseases. Arterio-sclerosis is very rarely a cause of spinal hæmorrhage.

Pathology.—The membranes and outer surface of the cord appear normal if the vertebræ are not injured. In primary hæmatomyelia the hæmorrhage nearly always begins in the grey matter, it is usually limited to the grey matter, and the lateral white columns of the cord are nearly always spared (*see* Plate IV, p. 392). The grey substance is more vascular and it is less firm than the white : also the largest spinal arteries pass to the grey matter, and in the grey matter the vessels are not surrounded by so much supporting connective tissue as in the white matter. These facts explain the localisation of the hæmorrhage.

The blood extravasation at the seat of the lesion may be so great that the blood escapes on section and a cavity is left, or it may be scattered over the transverse section in small patches—disseminated hæmatomyelia.

The hæmorrhage may invade the whole of the grey substance or one half of it only ; it may be limited to the posterior or anterior horn ; or it may be situated chiefly around the central canal (tubular haemorrhage) or in the ventral half of the posterior columns. With the exception of the part just mentioned, the hæmorrhage seldom affects the white matter, and then only in small patches as a rule. The hæmatomyelia which affects chiefly the central part of the cord is known as central hæmatomyelia.

The hæmorrhage often extends upwards and downwards in the grey matter and there is little tendency to spread laterally. Sometimes the hæmorrhage spreads throughout a great vertical extent of the cord —longitudinal central hæmatomyelia.

Goldscheider and Flatau have found, by the injection of coloured fluids into the cord post-mortem, that the injection fluid spreads longitudinally in the grey matter, especially in the posterior horns, much more readily than in the white. The white substance resists the extension of fluid either laterally or longitudinally, but the resistance is least in the ventral parts of the posterior columns.

The blood extravasation has a special tendency to extend longitudinally in the posterior horns : it has little tendency to extend within the central canal.

The cervical and lumbar enlargements of the cord, especially the former, are more frequently the seat of hæmorrhage than the dorsal region.

When the commissure is not destroyed mechanically, the blood extends independently in each half of the cord.

Minor has described a form of hæmorrhage in the grey substance, or ventral part of the posterior columns, in which the blood extravasation forms a ring or tube—hæmatomyelia annularis.

Around a spinal hæmorrhage myelitis is very liable to develop. The blood is usually absorbed by the end of six weeks (Minor), but it may sometimes be detected for a longer period. At the seat of the

hæmorrhage a cicatrix, or a cyst-like, round or oval cavity may form in course of time in the posterior columns, or a fissure may form in the posterior horns. Some of these cavities closely resemble those of syringomyelia.

Minute or capillary hæmorrhages ("accessory hæmorrhages") often accompany spinal diseases (myelitis, softening, etc.): also they occur in many infectious diseases. But clinically they are of little importance. Long vertical fissures in the posterior horns are often the result of old hæmorrhage.

The **symptoms** of hæmatomyelia are those of motor and sensory spinal paralysis of sudden onset: they usually become well marked in the course of a few minutes: but occasionally there is an increase of the paralytic symptoms, for an hour or two after their onset. There are usually no premonitory symptoms and consciousness is not lost as a rule. The patient has been apparently in good health up to the onset of the hæmorrhage.

The table on p. 109 indicates the mode of onset in various spinal diseases and shows the other affection in which the onset is abrupt.

In many cases the symptoms are those of a transverse lesion of the cord, which develop in a few minutes, viz.: motor paralysis and anæsthesia up to the level of the lesion, with paralysis of the bladder and rectum. The legs are flaccid at first and the knee-jerks lost. When the lesion is in the lumbar region these symptoms persist, and the leg muscles atrophy and present the reaction of degeneration.

When the lesion is in the dorsal or cervical regions, usually the knee-jerks soon return and become increased, ankle-clonus and the Babinski or extensor type of plantar reflex develop, and the legs become spastic. In cervical hæmorrhage, usually there is atrophic paralysis of the arm muscles and spastic paralysis of the leg muscles, as in a transverse cervical myelitis. Oculo-pupillary changes occur when the lesion is in the lowest cervical region—contraction of the pupil and diminution of the palpebral fissure (unilateral or bilateral).

As in all transverse lesions of the cord, when the destruction of the nervous element is total, the reflexes below the lesion may be permanently lost and the legs flaccid.

The sensory symptoms in a severe form of hæmorrhage are anæsthesia to tactile impressions, analgesia, and thermo-anæsthesia in the parts below the level of the lesion, with a band of hyperæsthesia (in some cases) at the level of the hæmorrhage. But in many cases the sensory affection has been loss of sensation to pain and temperature in the parts below the hæmorrhage without affection of tactile sensation (dissociated anæsthesia). These are the sensory symptoms so common in syringomyelia, and this dissociated anæsthesia is due to the hæmorrhage affecting only the grey matter.

Sometimes the hæmorrhage is entirely or chiefly unilateral, and then the symptoms of hemiparaplegia (or Brown-Séquard's paralysis)

are met with—paralysis of one leg with loss of sensation of the opposite side. But often the sensory affection has consisted of unilateral analgesia and thermo-anæsthesia, whilst tactile sensation has been preserved. In some cases when the hæmorrhage has been unilateral and situated in the cervical region, there has been atrophic paralysis of the arm and spastic paralysis of the leg on the side of the lesion, with loss of sensation to pain and temperature on the side opposite to the lesion, whilst tactile sensation has been preserved on both sides.

Minor has pointed out, that above the region of total anæsthesia in spinal hæmorrhage there may be a zone of analgesia and thermo-anæsthesia. This he regards as an indication of central hæmatomyelia.

The temperature is normal at the onset; but in a few days there is often a slight elevation of temperature, owing to the development of myelitis around the hæmorrhage.

At the onset of the hæmorrhage there is often pain in the back, but this is not invariably present; it is not usually severe, and it is only temporary. The pain is limited in extent, it is localised near the seat of the lesion, and may radiate in the nerve roots from the part of the cord affected by the hæmorrhage.

The paralytic symptoms due to the hæmorrhage often reach their highest development directly after the onset of the affection; but there is sometimes an increase of the paralysis at a later period owing to the development of secondary myelitis around the blood-clot. Vaso-motor disturbances, and trophic changes in the skin may develop.

Cystitis and bed-sores are two frequent and serious complications, as in myelitis.

Prognosis and Course.—After the first week improvement often occurs: the sensory symptoms diminish, and some of the paralysed muscles recover. Death is rare at the early period, though it does occur occasionally from paralysis of the respiratory muscles, when the lesion affects the cervical region. At a later period, death may occur from cystitis or bed-sores as in cases of myelitis. As regards danger to life at the early period, the prognosis is favourable. In course of time improvement usually occurs and often it is considerable; but complete recovery is exceptional.

Diagnosis.—The important point in the diagnosis is the very sudden onset of motor and sensory spinal paralysis. Without the occurrence of premonitory symptoms, the patient becomes completely paralysed in a few minutes' time. If premonitory symptoms have been present for more than a few minutes before the paralysis, the case is more probably due to myelitis than hæmorrhage. A history of injury or strain just before the onset of the paralysis would be in favour of hæmorrhage; whilst onset of paralysis at night would be against hæmorrhage.

The pain in the back in spinal hæmorrhage is limited to one region. In meningeal hæmorrhage the pain in the back is more severe and more extensive; it radiates into the limbs, and the back is rigid.

In myelitis and spinal softening the onset is not so sudden; usually there are premonitory symptoms (paræsthesia, etc.); a day or two elapse before the paralysis reaches its height; and the temperature is a little elevated at the onset. When the symptoms come on without apparent cause, myelitis is more probable than hæmorrhage.

There is one variety of myelitis—myelitis-apoplectica—in which the onset is practically as abrupt as in hæmorrhage; and the diagnosis of these cases is almost impossible (*see* p. 130).

In acute anterior poliomyelitis there is fever at the onset; the symptoms are entirely motor, and there is no anæsthesia to tactile impression, pain or temperature (unless the posterior grey matter is affected); also the bladder and rectum are not paralysed.

In central hæmatomyelia, as already mentioned, there is often loss of sensation to pain and temperature, whilst tactile sensation is preserved, and hence these cases resemble syringomyelia in their sensory symptoms; but in hæmatomyelia the onset of the affection is very sudden, and in course of time there is a tendency for gradual improvement, whilst in syringomyelia the onset is usually very gradual and the symptoms in course of time steadily increase.

Treatment.—At the early period, absolute rest is of the greatest importance. The patient should be kept in bed lying on the side or in the prone position. It is important that everything should be done to check coughing and to avoid straining in any way. In a strong individual leeches may be applied to the back, over the region of the hæmorrhage, or cupping at this spot may be employed.

An icebag applied to the spine over the affected part, is believed to be of service. The bowels should be kept freely open. Ergot (in the form of the liquid extract) or ergotin may be given at the onset; also ergotin may be injected subcutaneously.

After the early period has passed by, the treatment is the same as that of a transverse myelitis. The condition of the bladder should be attended to, and the urine should be drawn off with a catheter if necessary. Everything should be done to prevent cystitis, and if this symptom should develop, the bladder should be washed out with a mild antiseptic (*see* p. 138).

Care should be taken to prevent the formation of a bed-sore and the use of a water bed is advisable (*see* p. 137).

If the paralysed limbs are flaccid, the use of the galvanic current may be of slight service (*see* p. 139).

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MENINGEAL HÆMORRHAGE [1]

Meningeal hæmorrhage may occur either between the vertebræ and dura mater (extra-meningeal), or inside the dura mater (intra-meningeal). In the latter form the hæmorrhage may be in the arachnoid sac or in the pia mater.

In the extra-meningeal form, the hæmorrhage occurs from rupture of a vessel in the venous plexus between the dura and the vertebræ. Usually the haemorrhage is situated on the posterior surface of the dura and is not of great extent. The causes are chiefly traumatic—fracture of a vertebra, wounds, concussion, etc. Occasionally the extra-dural hæmorrhage has been due to tetanus, or to some disease causing violent convulsive attacks, or to the bursting of an aneurism into the vertebral canal.

The blood is seldom so abundant as to compress the cord ; but when an aneurism bursts into the vertebral canal, symptoms of compression "myelitis" are produced.

Intra-meningeal hæmorrhage is occasionally due to tetanus and eclampsia, and in children, at birth, it may be caused by physical conditions produced by difficulties in delivery. It occasionally occurs in diseases in which there is a hæmorrhagic tendency, as in scurvy and purpura : and also at the onset of very severe and acute meningitis.

Meningeal hæmorrhage may result, in rarer cases, from the extension of intra-cranial hæmorrhage into the spinal meninges. In these forms the symptoms are usually not well marked, and they are masked by those of the primary diseases.

Clinically, the most important form of meningeal hæmorrhage is the non-traumatic meningeal apoplexy in the arachnoid sac. The dural sac is filled with blood at the region of 2 or 3 vertebræ ; in other cases the greater part, or the whole, of the dural sac of the cord is filled with blood. The etiology of this form has not been clearly made out, but the most important cause is said to be muscular over-strain.

The chief **symptoms** of extensive meningeal hæmorrhage are those of sudden irritation of the meninges and nerve roots, followed by symptoms of spinal compression. The symptoms come on quite suddenly, the first being exceedingly severe pain in the back. Often the pain extends over the greater part of the spinal column. Frequently it is very severe in the sacral region. The pain extends along the course of the nerve roots irritated by the hæmorrhage and is associated with hyperæsthesia and paræsthesia. There is rigidity of the back, caused by pain and by muscular spasm : also there is spasm of the muscles supplied by the nerve roots at the seat of the hæmorrhage and rigidity of the muscles below the lesion. Soon paresis and diminution of sensa-

[1] Meningeal hæmorrhage is considered here rather than in the section devoted to diseases of the meninges, owing to the similarity of symptoms as regards sudden onset with those of hæmatomyelia.

tion develop in the parts below the lesion and paraplegia and anæsthesia may finally occur. Difficulty in passing urine and retention of urine often develop. At the onset there may be erection of the penis.

At first the tendon reflexes are diminished; later the reflexes may be increased below the lesion, as in hæmatomyelia.

As a rule, there is no loss of consciousness at the onset, but occasionally there has been unconsciousness, as a result of shock, for a short time.

Usually the symptoms reach their height in a few hours, and after complete rest there is a little improvement. Then from two to four days after the onset, the symptoms often increase owing to the development of secondary inflammatory changes.

Prognosis.—Death may occur within a few hours from shock, or from the hæmorrhage extending into the cranial cavity. After the first week there is little danger to life. Recovery, complete or almost complete, may occur, but cystitis and bed-sores are serious complications. In meningeal hæmorrhage in the cervical region there is danger of death from respiratory paralysis.

The **diagnosis** is based on the very sudden pain in the back with signs of irritation of the membranes and nerve roots.

From hæmatomyelia, the meningeal hæmorrhage can be diagnosed by the very severe and extensive symptoms of nerve root irritation in the latter disease. In hæmatomyelia, pain is more frequently absent, it is less intense, and not so extensive, also paralysis is present from the onset; whilst in meningeal hæmorrhage the paralysis is preceded by pain and symptoms of irritation of nerve roots and meninges. Dissociated anæsthesia (loss of sensation to pain and temperature whilst tactile sensation is normal) is often present in hæmatomyelia, but is absent in meningeal hæmorrhage.

In acute meningitis and meningomyelitis there is fever at the first and the onset of symptoms is not so sudden.

Treatment.—Absolute rest is necessary, best in the prone position or on the side.

Local venesection, by cupping or by leeches, is advisable. An icebag may be afterwards applied to the spine, at the part where the hæmorrhage is diagnosed, and ergot or ergotin may be given.

The bowels should be moved freely. For the severe pain morphia may be required. The late symptoms and complications require the same treatment as in myelitis.

Operative treatment, laminectomy and removal of blood-clot, has been recommended by several writers in cases where the hæmorrhage is extensive and causes danger to life, but its value has not yet been clearly demonstrated.

REFERENCE.

Leyden and Goldscheider. *Die Erkrankungen des Rückenmarkes.* Wien, 1897.

CAISSON DISEASE: DIVERS' PARALYSIS.

(*Paralysis from diminished atmospheric pressure.*)

A caisson is a metal cylinder or air chamber used under water in the construction of bridges. The caisson is fixed vertically, it is open at the lower end, which rests on the bed of the river, and the water is expelled from the caisson by compressed air. At the upper end are chambers or "locks," which can be hermetically closed. Workmen enter the caisson through these "locks," and do their work in a greatly compressed atmosphere.

Divers work under similar conditions, owing to the increased pressure of the atmosphere in their air-filled helmets.

Divers and men who work in caissons sometimes suffer from spinal and cerebral symptoms on returning to the surface (i.e. to a normal atmospheric pressure), especially if the change should be made suddenly. The cerebral symptoms are usually slight and transient: they consist chiefly of headache, faintness, giddiness, nausea, vomiting, temporary mental disturbances, pain in the ears and deafness, epistaxis, coma of short duration, and occasionally temporary hemiplegia or monoplegia.

The spinal symptoms are more common, and usually more severe than the cerebral. Soon after returning to the surface, or, it may be in about half an hour or later, there are severe pains in the legs and trunk, and occasionally in the arms. These pains are referred to the large joints (knees and elbows), but there is no swelling in the joints. Soon afterwards, the legs become weak and heavy, and feel numb. Paralysis of both the legs develops rapidly (often in a few minutes), and soon there is complete paraplegia. The symptoms are usually those of a transverse lesion of the cord, and spastic paraplegia develops.

Parasthesia and anæsthesia may also develop, but the latter is often incomplete or irregular in distribution. The sphincters are affected in severe cases. The arms are rarely affected. In some cases, the symptoms are chiefly ataxia, with sensory symptoms.

In mild cases, recovery occurs in a few days; in other cases in a few weeks or months; whilst in severe forms the paralysis may be permanent.

In a few cases, death occurs suddenly, with or without delirium and convulsions.

Etiology and Pathology.—The patient has usually worked at 40 to 90 feet below the surface of the water, and at a pressure of 2 to 4 atmospheres.

The paralysis does not occur whilst the men are working in the caisson or under the water, but when they return to the ordinary atmospheric pressure on the surface.

A very important factor in the causation of the paralysis is the *sudden* return to the normal atmospheric pressure; also the symptoms are more likely to occur the higher the atmospheric pressure in the caisson,

and the longer the hours of work at this high pressure. Healthy temperate men are less liable to suffer than alcoholic individuals. Those who have been employed in the work for some time are less liable to suffer than those recently engaged.

The **pathological anatomy** has been studied in a few cases.

In a case reported by Leyden, in which death occurred at the end of fifteen days, pathological changes were found in the dorsal region of the cord, in the posterior columns, and in the posterior part of the lateral columns. The cervical and lumbar regions were normal. At the part affected fissures were seen in the nerve tissues, and these fissures contained large round nucleated cells. The nervous tissues around the fissures presented the appearance of parenchymatous myelitis and the nerve fibres were swollen. In the anterior columns and in the anterior part of the lateral columns little change could be detected. The grey substance, the nerve roots, and the meninges were normal. The pathological changes were symmetrically distributed and there were no signs of hæmorrhage.

In a case recorded by Schultze death occurred at the end of 2½ months. The cord presented disseminated myelitis in the dorsal region. In other cases multiple necrotic patches have been found in the lower dorsal region.

A review of the pathological anatomy of the cases on record shows, that in recent cases empty spaces are found in the neuroglia network, or spaces containing swollen axis-cylinders surrounded by a narrow rim of myelin. The blood vessels and their perivascular lymph sheaths are dilated ; and scattered small hæmorrhages may be present. In the grey substance, the pericellular spaces are dilated, the nerve cells granular and vacuolated, and their nuclei atrophied and badly stained. At a later stage clusters of granular cells are found in spaces in the nerve tissues, and there are patches in which the tissues have degenerated completely and only detritus is left in place thereof. There is no cell infiltration of the tissue.

The changes closely resemble those seen in dogs in which the disease has been produced experimentally. The pathological and experimental study of the subject, as well as the etiological facts already mentioned, strongly support the view, that the paraplegia and other symptoms of the disease are caused through the rapid escape of gas from the blood when the workman returns suddenly to the ordinary atmospheric pressure. The gas liberated causes rupture of the nervous tissues, and fissures are formed ; in the capillaries, it obstructs the circulation acting as air emboli, and as a result there is ischæmic softening of the tissues supplied by the obstructed small vessels. In man the symptoms are not caused by spinal hæmorrhage. The changes chiefly occur in the dorsal region, which is the least resisting part of the cord, and in the region supplied by the postero-lateral arterial system—the posterior and lateral columns ; whilst the most vascular part of the transverse cord section (supplied by the anterior median arteries) escapes.

The gas which escapes from the blood is believed to be nitrogen by some writers, by others oxygen and carbonic acid.

During the time spent in the compressed atmosphere the blood becomes charged with gases from the air. On the sudden return to the normal atmospheric pressure these gases escape rapidly and rupture the tissues, or cause air emboli and softening as already stated.

Experiments on animals have shown, that when the animal has been subjected to a high atmospheric pressure and then suddenly been brought into the normal atmosphere, gas bubbles are seen in the blood vessels and in the parenchyma of the spinal cord. Hill and McLeod have demonstrated these bubbles of gas in the capillaries in the web of the frog's foot and the bat's wing. The air emboli could be seen arresting the circulation in the small vessels. On "recompressing" the animal the air emboli were absorbed and the circulation was re-established. When "decompression" of the animal was effected slowly no gas bubbles appeared.

Oxygen at a high tension has a toxic action on the nervous system, and this has been regarded by Prof. Lorrain Smith as an important factor in the causation of the disease.

Prognosis.—Fortunately severe forms of the disease are rare; death occurred in 12 out of 76 cases at the building of the St. Louis bridge, and in 3 out of 110 cases at the construction of the Brooklyn bridge.

The chief dangers to life, after the first few days, are the complications, such as those which occur in transverse myelitis.

The **diagnosis** is easy, and is based on the history of paraplegia developing in a man who has been working in a caisson or as a diver, directly after a sudden return to the ordinary atmospheric pressure.

Treatment.—Directly the symptoms of the affection develop the man should be subjected to recompression, i.e. he should return to the caisson or to a pneumatic cabinet in which a high atmospheric pressure can be produced immediately. (Later the return to the normal atmospheric pressure must be very gradual.) This treatment relieves the pains in the limbs and other symptoms. For the pain morphia is sometimes necessary. Ergot has been recommended in frequent and large doses at the onset. The late treatment is the same as that of myelitis and its complications.

In order to prevent the occurrence of the disease, it is important that the return to the ordinary atmospheric pressure should be very gradual. The man should enter the lock connected with the caisson and the atmospheric pressure should be very gradually diminished. The hours of labour should not be too long. Only healthy and temperate men should be engaged for the work. The caisson should be very thoroughly ventilated.

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Pick. Article, *Handbuch der pathologischen Anatomie des Nervensystems*. Berlin, 1903.

SECTION VIII.

DISEASE OF THE CORD CAUSING ATROPHIC PARALYSIS

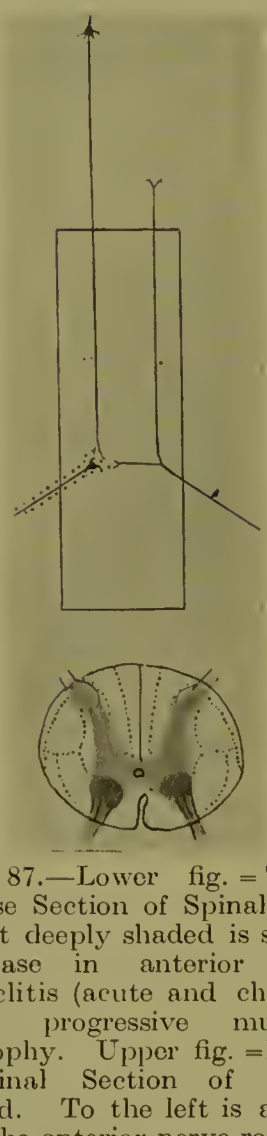


FIG. 87.—Lower fig. = Transverse Section of Spinal Cord. Part deeply shaded is seat of disease in anterior poliomyelitis (acute and chronic), and progressive muscular atrophy. Upper fig. = Longitudinal Section of Spinal Cord. To the left is a fibre of the anterior nerve root and its cell (lower motor neuron). Above it is the upper motor neuron. To the right is the posterior root and ascending fibre—sensory neuron. In the diseases just mentioned the lesion affects only the lower motor neuron—marked with dots in the fig.

WE have now to consider the disease of the cord in which atrophic paralysis is the prominent feature.

The various diseases causing atrophic paralysis are given in the following table.

DISEASES CAUSING ATROPIC PARALYSIS.

I. SPINAL LESIONS.
- Acute anterior poliomyelitis of the infant (infantile paralysis).
- Acute anterior poliomyelitis of the adult.
- Subacute anterior poliomyelitis.
- Chronic anterior poliomyelitis.
- Progressive muscular atrophy.
- Amyotrophic lateral sclerosis.
- Hereditary progressive spinal muscular atrophy of childhood (Hoffmann and Werding).
- Peroneal type of muscular atrophy (Charcot, Marie, Tooth).
- Syringomyelia.
- Cervical pachymeningitis.
- (Various lesions affecting the anterior horns of grey matter of the cervical or lumbar region may produce atrophy of muscles supplied from the diseased horn, as in—
  Spinal hæmorrhage, myelitis, compression myelitis from caries or tumour of the vertebræ, tumour of the cord or its meninges.)

II. NERVE LESIONS.
- Injuries and diseases of nerves.
- Various forms of peripheral neuritis, multiple and localised.

III. MUSCLE LESIONS.
- Muscular dystrophy
  - A. Pseudo-hypertrophic paralysis.
  - B. Idiopathic muscular atrophy.
    - (a) Scapulo-humeral type (Erb's juvenile type).
    - (b) Facio-scapulo-humeralor infantile type (Landouzy-Dejerine).

(The affections in Groups II and III do not come within the subject of this work.)

The diagram, Fig. 87, indicates the position of the lesion in those diseases, named in the table, which affect only the lower motor neurons.

ACUTE ANTERIOR POLIOMYELITIS: INFANTILE PARALYSIS AND ACUTE ATROPHIC PARALYSIS OF THE ADULT

(πόλιός = grey)

Acute anterior poliomyelitis is a disease characterised clinically by paralysis of the muscles of one or more limbs, which develops rapidly, and is followed by muscular wasting, but is unaccompanied by anæsthesia. The disease is usually met with in children; but it occurs occasionally in the adult (anterior poliomyelitis of the adult).

The disease may begin (1) with, or (2) without febrile symptoms. In the cases with a febrile onset the temperature rises to 101° or 102° F., and may remain about 101° F. for a few days. There is often headache, drowsiness, and loss of appetite: convulsions and vomiting occasionally occur. There is often slight pain and sometimes great tenderness in the limbs. The febrile symptoms continue for a few hours or a few days. Within one or two days of the onset of the febrile symptoms paralysis occurs rather rapidly. Often the child is put to bed with febrile symptoms, and is found to be paralysed next morning; sometimes the paralysis is not noticed for a few days, if the child be kept in bed. There is sometimes pain in the joints, back and neck, and pain when the limbs are moved. In rare cases there is slight rigidity of the back and neck, suggestive of slight meningitis. Temporary cerebral symptoms occasionally occur at the onset, such as somnolence, occasional diplopia, slight delirium, and giddiness; but these symptoms soon pass away.

In many cases febrile symptoms, pain and tenderness, and signs of ill health are entirely absent, and the paralysis comes on suddenly, like that of spinal hæmorrhage. Allen Starr found that in 100 consecutive cases 69 began with fever, 31 without febrile symptoms. He believes that the two modes of onset probably indicate a difference in the exact nature of the pathological lesions.

The **symptoms** of infantile paralysis are chiefly those of motor paralysis of rapid onset without anæsthesia. Often there is no increase of the paralysis after it is first noted; in other cases it extends rapidly for a short time, but after the first few days there is no further increase. The early paralysis is more extensive than that which remains permanently. The paralysis at first may affect both legs or all four limbs, or three limbs, or one leg and one arm (of the same or opposite sides), or all, or a portion, of the muscles of one limb. In course of time great improvement may occur, and only one limb or one group of muscles may remain permanently paralysed. The improvement in the paralysis is slow, and commences from one to eight weeks

after the onset. It is first noticed in the muscles which were least affected at the commencement of the disease. It is only after three months that any opinion can be formed as to the extent of the permanent paralysis, and even after that time there is improvement; but complete recovery very rarely occurs.

The following table shows the distribution of the permanent paralysis in 595 cases. It is compiled by Allen Starr, from his own cases and those of Duchenne, Seeligmüller, and Sinkler :—

One leg (right, 123 ; left, 123)	246
Both legs	170
One arm (right, 26 ; left, 21)	47
Both arms	6
All limbs	47
Arm and leg of same side	33
Arm and leg of opposite sides	8
Trunk	26
Three extremities	12

Occasionally the muscles of the back are paralysed. In rare instances muscles suppled by the seventh, twelfth, third, or sixth cranial nerves have been paralysed ; but this has been almost always in epidemics of the disease, and has been due to the extension of the pathological changes to the medulla.

In a large number of cases of infantile paralysis only a portion of the mucles of a limb, or one group of muscles, or occasionally a single muscle, remains permanently paralysed.

Paralysis of one leg is the most common form of infantile paralysis. There are two types—the leg type and the thigh type. In the former the anterior tibial and peronei muscles only, or one or two muscles of this group, are permanently paralysed. Occasionally only the calf muscles are paralysed. In the thigh type, the psoas and iliacus, the glutei, and the thigh muscles are affected only. Often the sartorius is spared.

In the arm, the deltoid, biceps, and supinator longus may be paralysed together, or the deltoid may be alone affected. In rare cases, the forearm muscles or the small muscles of the hand are chiefly affected.

The paralysed muscles not being able to perform their functions, defects in the movements of the limb occur. Thus, when only the anterior tibial muscles are paralysed there is dropping of the foot and of the toes ; the patient cannot dorsiflex the foot nor extend the toes, but he can perform the movements in the opposite direction (flexion of the toes and plantar flexion of the foot). In walking there is dropping of the foot on the affected side, and the leg is raised unusually high at the knee at each step, to clear the toes from the ground.

The paralysed muscles are flaccid ; those which remain permanently paralysed undergo atrophy rapidly, hence the name acute atrophic paralysis.

Electrical examination reveals changes in the reactions by the end

of the first week if not before. The changes vary from slight altered excitability up to partial or complete reaction of degeneration. In the latter condition there is loss of the excitability of both the muscles and their motor nerves to the faradic current. To galvanism there is no response in the nerves, but in the muscles the excitability is at first increased for a few months ; the anodal closing contraction is often greater and more readily obtained than the kathodal closing contraction, and the jerk of the muscle is more sluggish than in health. At a later period, in the muscles which remain permanently paralysed, there may be no response to either faradism or galvanism. At the onset it is noted that some of the paralysed muscles react to faradism, whilst others (presenting the reaction of degeneration) do not. The paralysed muscles in which the faradic contractility is not lost within the first week will ultimately recover.

The bladder and rectum are not paralysed, and bed-sores do not develop.

The reflexes are lost when the cord lesion destroys the " reflex arc " on the integrity of which the reflex depends, or when muscles which perform the reflex movement are paralysed. Thus, when one leg is paralysed, the knee-jerk is often absent on the paralysed side, but is present in the other non-paralysed leg. When the thigh muscles are paralysed the knee-jerk is always absent. When the anterior tibial muscles only are paralysed the knee-jerk may be absent at first, but at a later period it may be present. When the calf muscles are paralysed the tendo Achillis reflex is absent. If muscles are paralysed which are normally thrown into action in the " superficial reflexes," the reflex movement cannot be produced. Thus, if the muscles of the foot and toes are paralysed they do not contract when the sole of the foot is irritated ; only the muscles of the hip and the flexors of the knee are then thrown into action.

There is no anæsthesia in a true poliomyelitis limited to the anterior horn of grey matter ; but Allen Starr states that he has so frequently noted a permanent increased sensibility to pain in the paralysed limb that he " cannot but believe that the lesion in the grey matter affects the pain-sense tracts in their passage through the cord at the level of their entrance." In a case which I have recorded there was unilateral loss of sensation to pain and temperature, whilst tactile sensation was normal ; but in this case microscopical examination showed that the poliomyelitis extended into the posterior horn of grey matter on the side on which the sensory affection was noted.

Condition at a Late Period.—If many muscles of a limb have been paralysed, then the limb will remain useless ; often, however, there is slow recovery, even at a late stage, and the muscles of a limb which have escaped paralysis supplement, to some extent, the action of those paralysed.

In course of time the paralysed limb is generally much wasted, and

the girth and length are distinctly less than those of the opposite limb, if the latter should be unaffected.

Occasionally the muscular wasting is obscured by a development of interstitial fat.

In course of time the paralysed limb becomes bluish and livid. The circulation is feeble, and the limb feels colder than the opposite healthy limb.

Owing to paralysis of certain groups of muscles, whilst others are

FIG. 88.—Infantile Paralysis. Dropped foot. Paralysis of anterior tibial muscles. Foot in position of talipes equinus.

FIG. 89.—Condition of both feet similar to that in Fig. 88. Old infantile paralysis.

not paralysed, various deformities are produced by the over-action of the latter. Talipes equinus or equino-varus is the most common deformity; it is due to the paralysis of the anterior tibial group and peronei, and over-action of the normal opponents; also the action of gravity favours the production of this deformity. Talipes varus

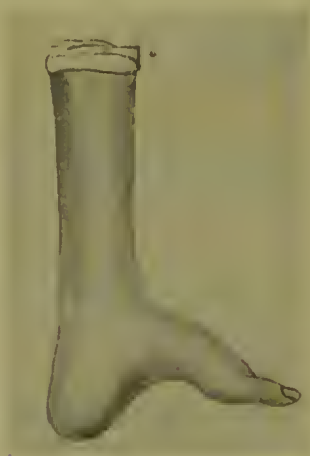


FIG. 90.—Old Infantile Paralysis. Calf muscles paralysed and atrophied. Pes calcaneus—heel projecting.

FIG. 91.—Paralysis of Left Deltoid (to the right of the photograph). Note wasting of muscle and slight depression under outer end of clavicle.

is due to paralysis of the peronei; talipes valgus to paralysis of the tibialis anticus; talipes calcaneus to paralysis of the calf muscles. Pes planus is caused by paralysis of the peronei and plantar flexors. Lateral curvature of the spine often occurs when one leg is paralysed.

The ligaments of the joints become lax, and dislocations and subluxations are apt to occur. Genu valgum is occasionally met with. Occasionally the knee assumes a flexed position. In course of time a limb may become fixed in one position, through contraction and fibrous degeneration of the muscular tissue of an opponent of a paralysed muscle. Bones become arrested in their development in the paralysed limb, and are found to be smaller and shorter than those on the non-paralysed side.

Fig. 92.—Atrophy of small Muscles of the Hand (thenar and hypothenar). Old infantile paralysis.

Prognosis. —The disease is seldom fatal. Death occurs in rare cases, when there are marked febrile symptoms and extensive paralysis at the onset. Complications at this stage and paralysis of the respiratory muscles occasionally cause a fatal termination.

Usually improvement occurs. Many of the muscles at first paralysed recover in course of time. Even after a long period of paralysis some muscles recover ; but nearly always one or more muscles remain permanently paralysed. According to Wichmann complete recovery sometimes occurs : and in older children and adults the prognosis is worse than in young children.

Allen Starr thinks the prognosis is much better in cases which begin with fever than in those without a febrile onset.

Paralysed muscles which at the end of two or more weeks present only a slight change in the electrical excitability, or a partial reaction of degeneration (i.e. muscles which retain their faradic excitability), will gradually recover in course of time. But the prognosis is much worse with respect to the paralysed muscles which at this time present a complete reaction of degeneration (i.e. which do not react to faradism) ; they will only recover after a long interval, six to nine months, or they will never recover.

It is not possible to say what duration of paralysis renders recovery out of the question ; but generally a muscle which has been paralysed for nine months will not recover.

When one leg is paralysed, but the other unaffected or only partially paralysed, the patient is usually able to get about with the aid of supports in course of time.

Occasionally infantile paralysis appears to cause a predisposition to atrophic paralysis in later life. Rare cases are on record in which chronic nervous affections, such as chronic anterior poliomyelitis, muscular dystrophy, diffuse cervical myelitis (producing symptoms like amyotrophic lateral sclerosis), and " professional " atrophic paresis, have developed in adult or middle life, in individuals who have suffered from acute anterior poliomyelitis in infancy.

The **diagnosis** of acute anterior poliomyelitis of the infant is usually

easy. The characteristic features of the disease are: The acute onset of motor paralysis, without anæsthesia and without affection of bladder or rectum; the rapid wasting of the muscles, with the reaction of degeneration. At the onset the paralysis may be *overlooked* for a time owing to the prominence of gastric and febrile symptoms. The latter may be regarded as the only abnormal conditions.

From *acute transverse myelitis* the diagnosis is usually easy. In poliomyelitis often only one limb is paralysed; but when both legs are affected the absence of anæsthesia, of affection of bladder and rectum, and of bed-sores would be points against myelitis.

Multiple *peripheral neuritis* is rare in children: it may be distinguished from acute anterior poliomyelitis by its more gradual onset, by the presence of sensory symptoms, pain in the limbs, tenderness of the muscles and nerve trunks, and hyperæsthesia, and by the symmetrical bilateral nature of the paralysis. Also, there is often a history of one of the well known causes of neuritis.

In *spinal hæmorrhage* there are sensory symptoms, loss of sensation to pain and temperature or to all forms of sensation, and often paralysis of bladder and rectum.

The forms of *progressive muscular atrophy* in children are distinguished by their gradual onset and the progressive increase of symptoms: whilst in acute anterior poliomyelitis the onset is sudden and there is a tendency for considerable improvement in course of time.

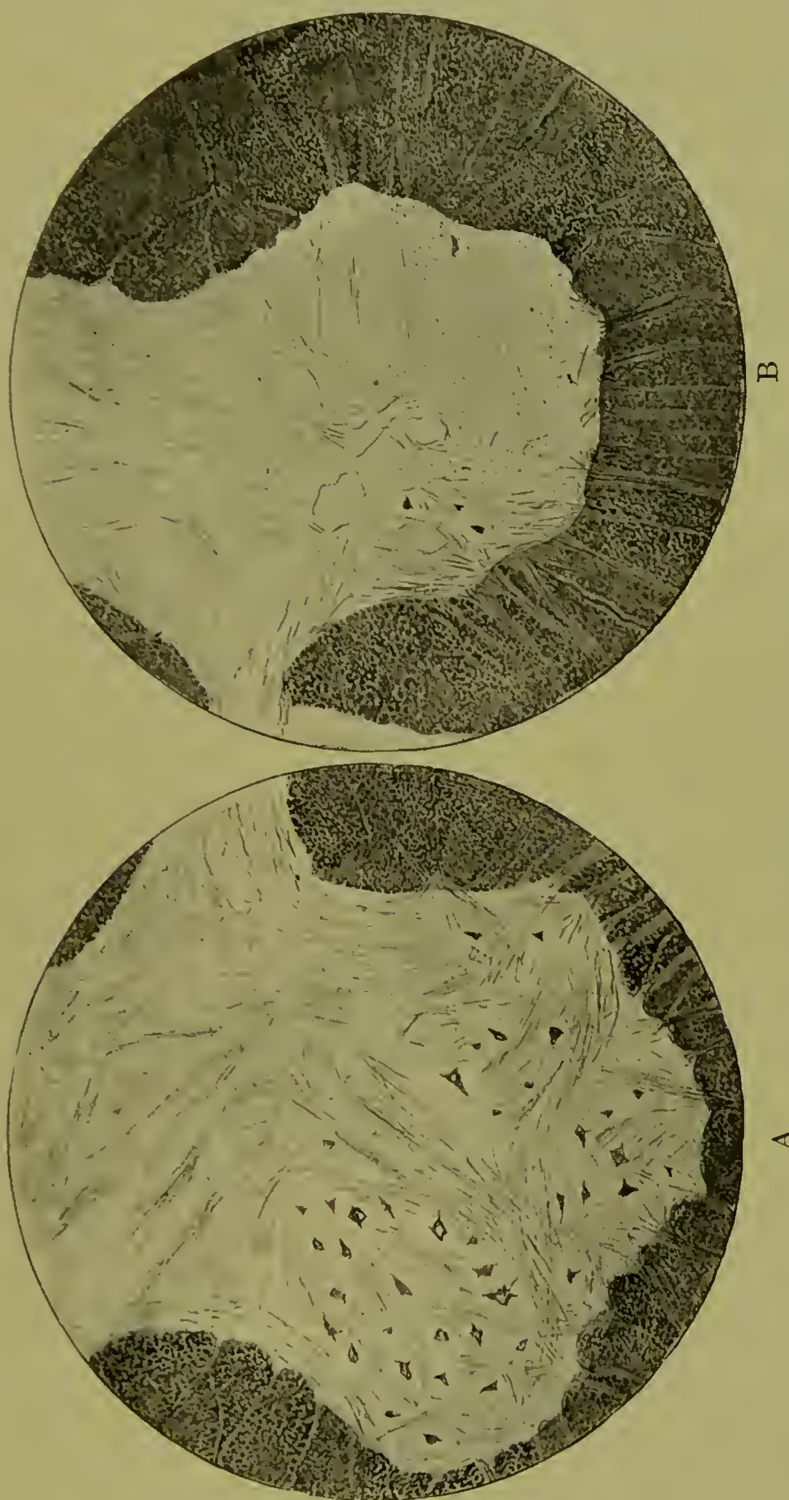
From infantile paralysis due to a *cerebral lesion*—spastic hemiplegia of infancy—the diagnosis is easy. In infantile spastic hemiplegia the paralysis is unilateral, there is rigidity of the limbs, increased reflexes and other signs of spastic condition, absence of the reaction of degeneration, and when atrophy is present it is diffuse and not localised to one group of muscles.

Erb's paralysis of the arm (paralysis of the deltoid, biceps, brachialis anticus and supinator longus) may be mistaken for infantile paralysis: but the history of the onset after injury, or its presence from birth and the history of difficult delivery, are important points: also sensory symptoms may be present, at the early stage. Diminished cutaneous sensibility may be then detected, in some cases, in the distribution of the 5th or 5th and 6th cervical nerve roots.

Pathological Anatomy.—As the disease is rarely fatal at an early stage, most of the pathological examinations have been made many years after the onset of the affection. It will be convenient to describe first the late changes, which are found years after the onset, and afterwards to consider the early changes.

Late Changes.—The paralysed *muscles* are greatly atrophied, and are of a greyish red, yellowish white, or rose colour. Microscopically, the muscle fibres are very narrow, the muscle substance is almost entirely absent, the muscle sheaths remain, and the muscle nuclei are markedly proliferated. Other muscle fibres are granular, and their transverse

striation is lost. In rare cases a single large muscle fibre may be seen amongst surrounding atrophied fibres. Occasionally there is a great increase of fibrous tissue and fat between the atrophied muscle fibres.



FIGS. 93 (A and B).—Spinal cord. Infantile paralysis. Late changes. Pal's stain. A = normal anterior horn of grey matter. B = anterior horn on side of paralysis. Note diseased anterior horn B is much smaller than normal horn A. The nerve cells and fine nerve fibres of grey matter in A are normal. In diseased horn B nerve cells have mostly disappeared, only a few remain at inner part: also fine nerve fibres are scanty; at outer part of horn B they are absent; here the nuclei of the neuroglia, represented by fine dots, are increased in number.

In extreme cases the degeneration of the muscle fibres is complete. The bones of the paralysed limbs are small and shorter, and their Haversian canals are smaller.

The *spinal cord*, when examined long after the onset of the disease,

presents well marked changes microscopically, though macroscopically little or no change may be detected. Usually the changes are unilateral,

FIG. 94.—Anterior half of Spinal Cord, thirty years after the onset of infantile paralysis. Weigert's stain. Note pale portion in outer part of anterior horn of grey matter to the right of the photograph. Nerve fibres and cells absent at this region; anterior horn to the left normal.

though the other side may not be quite normal. If one limb only has been paralysed (the usual form), the anterior horn of grey matter on the affected side is smaller than the other horn (*see* Figs. 93 A and B). Sometimes the whole of the half of the cord is a little smaller on the paralysed side. The changes, of course, occur in the lumbar region when the leg is affected, in the cervical region when the arm is paralysed. The nerve cells have disappeared or atrophied greatly, in the affected anterior horn, or in a portion thereof. Often degeneration is marked near a vessel, whilst the cells are normal a short distance from the vessel. In sections stained according to Weigert's or Pal's method the fine nerve fibres of the grey matter of the affected part of the anterior horn are seen

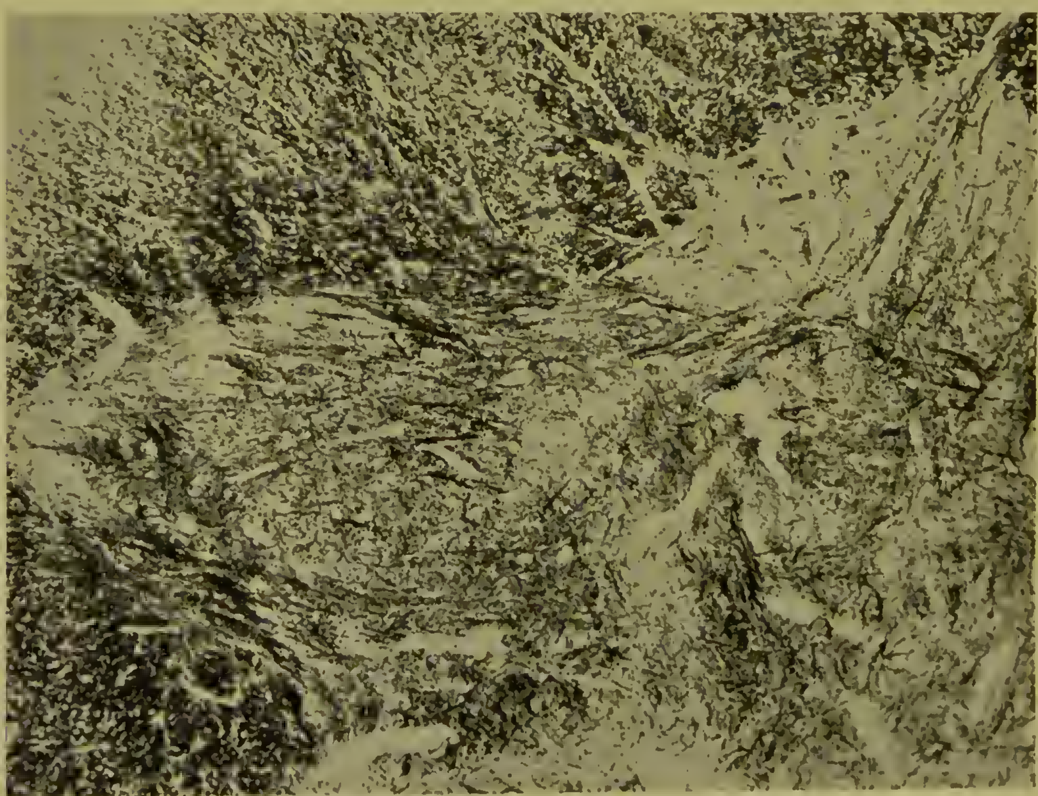


FIG. 95A.—Normal Anterior Horn of Grey Matter. Weigert's stain. For comparison with Fig. 95B. From same section as Fig. 94, but more highly magnified.

to be very scanty or almost entirely destroyed (*see* Figs. 93, 94, 95), whilst in the posterior horn they are normal. The neuroglia of the diseased anterior horn is increased in amount, especially around the vessels, and the neuroglia nuclei are slightly proliferated; spider cells are also present. The walls of the blood vessels in the affected anterior horn and in the anterior fissure are thickened in their external and middle coats. In

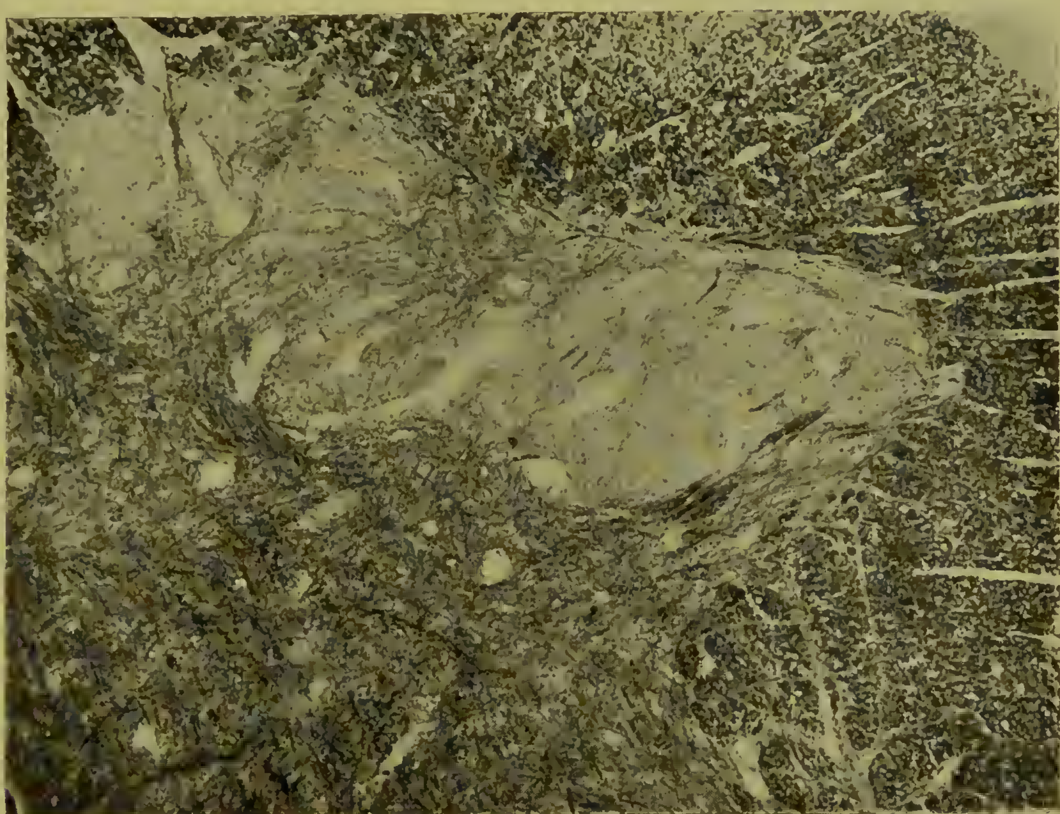


FIG. 95B.—Anterior Horn of Grey Matter of Spinal Cord. Old infantile paralysis. Note pale area from which nerve fibres and cells have almost disappeared. Weigert's stain. From same section as Fig. 94, but more highly magnified. Figs. 95A and B represent the anterior horns (normal and diseased) seen in Fig. 94.

some cases the changes in the neuroglia are absent, and there is simply atrophy and disappearance of the nerve cells and fine nerve fibres of the anterior horn, or of the nerve cells only. With the exception of the anterior horn of one side (or of both sides if the paralysis is bilateral) the rest of the grey matter is usually unaffected. But Schultze has pointed out that occasionally there is atrophy of nerve cells in Clarke's columns.

In many cases the changes affect only a portion of the anterior horn, as shown in Fig. 94.

The anterior commissure may show partial destruction of fibres near the affected horn; also there may be a diminution of nerve fibres in the white matter around the anterior horn.

The anterior nerve roots and the muscular branches of the peripheral nerves are atrophied.

In cases carefully investigated the atrophy of nerve cells has not been localised to definite groups of cells, but has followed, more or less, the distribution of small arteries in the anterior horn (Goldscheider).

The late changes in infantile paralysis are much less extensive than those at an early period. They are usually found to be limited to one or, occasionally, both anterior horns of the lumbar, sacral, or cervical region, according to the locality of the paralysis, and usually the vertical extent of the affection of the anterior horn is not more than 1 or 2 c.m.

In infantile paralysis, as already described, often certain muscles of a limb are paralysed, whilst the rest are spared. Spinal localisation, or the relation of various segments of the cord to the muscles of the limbs (as worked out by Ferrier, Thorburn, Starr, and others), explains in part the distribution of the paralysis. Thus, in the leg the tibialis anticus, extensor longus digitorum, and peronei are often paralysed, whilst the other muscles are spared. According to Thorburn these muscles receive their nerve supply from the second sacral segment; according to Kocher from the first segment. We may localise the lesion, therefore, to the level of the first or second sacral segment as regards its vertical extent. But other muscles supplied from this segment of the cord are spared. It appears probable, however, that the localisation of motor functions to various groups of nerve cells in the anterior horn (as worked out by A. Bruce, van Gehuchten, and others) furnishes the explanation of the limitation of the paralysis to certain combinations of muscles. According to A. Bruce the posterior lateral group of nerve cells of the anterior horn, at the level of the first and second sacral segments, supplies the motor fibres to the calf muscles and extensor muscles of the foot; whilst the nerve cells of the antero-lateral and central groups supply the gluteus medius and minimus, the external rotators of the hip, and the hamstring muscles. If the former group of nerve cells were chiefly affected and the latter spared by the lesion, this would explain the limitation of the paralysis to the anterior tibial group of muscles.

Mott and Bing have drawn attention to the degeneration in the white matter shown in sections stained according to Marchi's method. Degenerated fibres were seen in the anterior commissure, anterior ground bundle, and root zone; in the antero-lateral ascending tract and in the direct cerebellar tract to a less extent. In the posterior columns degenerated fibres were found in the parts containing the endogenous fibres. The degeneration in the posterior columns diminished from below upwards.

Early Changes.—When the pathological examination is made at an early stage of the disease the changes are, of course, very different from those just described.

Thrombosis of a vessel in the anterior horn was found by Angel Money many years ago. More recently Batten has recorded a case in which thrombosis was found in the same region along with cell

exudation and hæmorrhages. Batten draws the following conclusions from his pathological observations : (1) That the pathological changes in the anterior grey matter are due to a " primary thrombosis of a branch or branches of the anterior spinal artery supplying the grey matter of the anterior horns." (2) " That such thrombosis may be produced by many and various forms of infection and the disease is not due to a special specific infection." (3) " That the condition is more likely to occur in the lumbar region, owing to the blood supply of this portion of the cord being at a point more distant from the heart, and the long course of the reinforcing arteries."

Starr thinks it is probable that, in the case in which there are no febrile symptoms at the onset, the lesion is a primary degeneration of nerve cells (parenchymatous poliomyelitis), or it is due to hæmorrhage or to thrombosis in a small vessel in the anterior horn.

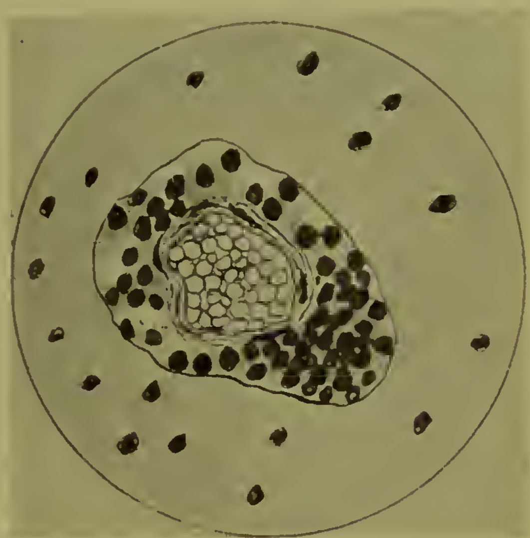


FIG. 96.—Section of Small Blood Vessel of Spinal Cord. Acute anterior poliomyelitis. Perivascular sheath distended and filled with nucleated cells (logwood stain).

But these views have been disputed. Certainly thrombosis or hæmorrhage have been very rarely found in the cases examined at an early period, and the cell infiltration around the vessels in the anterior median fissure before the vessels enter the cord, appears to be evidence against a *primary* thrombosis.

Some observers (Charcot and v. Kahlden) believe that in certain cases, at least, the initial change is an acute degeneration, limited entirely to the nerve cells of the anterior horn, i.e. that the lesion is sometimes an acute *parenchymatous* poliomyelitis. It must be allowed, that in many cases in which the pathological examination is made long after the onset of the disease interstitial changes are very slight, in comparison with the marked changes in the nerve cells.

But in nearly all of the very early cases examined within recent years the changes have been inflammatory in nature, i.e. the lesion has been an *interstitial* anterior poliomyelitis. The blood vessels of the anterior horn are seen to be dilated and full of blood. The perivascular lymph sheaths are crowded with round cells (*see* Fig. 96). The grey matter of the anterior horn is infiltrated with mono-nuclear round cells, compound granular cells, and with large cells possessing a large nucleus—so-called epithelioid cells (v. Leyden). Here and there are a few red corpuscles, and in severe cases there may be actual blood extravasation in the anterior horn. The cell infiltration is most marked around the vessels. The fine nerve

fibres in the grey matter are broken down where the cell infiltration is present. Sometimes the nerve cells are fairly well maintained in the centre of the cell infiltration; in other cases they are degenerated. They may be turbid, and their nuclei and processes indistinct; they are often swollen and granular; on staining according to Nissl's method the Nissl's granules cannot be detected, but the cell stains faintly in a diffuse manner. Finally the cells are atrophied or entirely broken up. Sometimes only one group of nerve cells is much affected; frequently only the external or median group of cells in the anterior horn is the seat of the lesion. But the changes are not specially localised to the cells of one or other group. In serial sections it is seen that the cells of a group which are spared at one level are affected at another level. Often some of the cells of one group degenerate whilst others are normal. It has been shown that in one group some of the cells are supplied by one small artery, whilst adjacent cells of the same group are supplied by another artery, and this explains why some cells are affected and others spared. The changes are often seen to follow the branches of a small artery.

Fig. 97.—Large Epithelial-like Cells (Leyden's) from diseased anterior horn in a case of acute anterior poliomyelitis.

The vessels in the anterior median fissure are often markedly altered. They are distended with blood, and their walls and the surrounding loose tissue are infiltrated with round cells. These cells increase as the vessels enter the anterior commissure. Sometimes the vessels entering the anterior horn from the periphery of the cord—anterior root arteries—are surrounded with round cells.

The chief changes in the cord are in the anterior horn; but sometimes the adjacent anterior white matter is also slightly affected. In exceptional cases the posterior columns have been slightly invaded by the myelitis. Also the changes in the grey matter occasionally extend to the base of the posterior horn, and rarely into the posterior horn itself. In a case which I have recorded the changes affected both anterior horns and intermediate portions of the grey matter, and on one side extended a little into the posterior horn (*see* Fig. 98).

Clarke's column is sometimes affected.

The pia mater of the anterior part of the cord is slightly or moderately infiltrated with cells, chiefly mono-nuclear leucocytes.

In some severe cases the changes in the grey matter affect the anterior horn on both sides of the cord in all regions, though they are best marked in the lumbar, sacral, and cervical regions, and the vessels of the pia mater and the anterior median arteries of all these regions of the cord have their walls infiltrated with round cells.

Changes similar to those described in the anterior horn have occa-

sionally been found in the motor nuclei of the medulla oblongata, but this is a very rare occurrence. Redlich has recorded the occurrence of poliomyelitis and encephalitis in the same patient.

Even in recent cases the paralysed muscles may present fatty degeneration. The nerve fibres of the anterior roots and the muscular branches of the peripheral nerves show well marked degeneration.

Pathogenesis.—In recent years numerous pathological examinations, made at an early stage of the disease, have shown that, in these cases at least, the affection is of vascular relation, and is one in which the

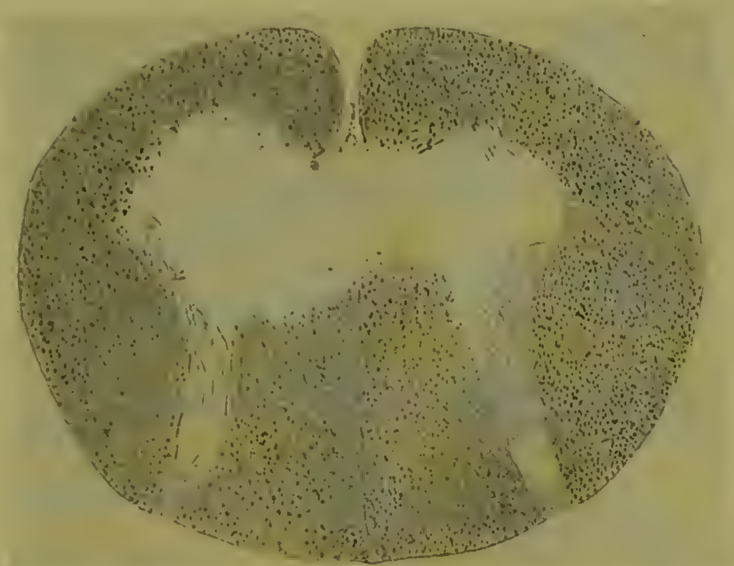


FIG. 98.—Section of cord in a case of acute anterior poliomyelitis. Early changes. Weigert's stain. Black dots = healthy nerve fibres; black lines = healthy fibres in grey matter. Pale blank part = area of myelitis. Area supplied by central artery of the cord chiefly affected. On the left-hand side of the diagram the poliomyelitis extends into the posterior horn, and posterior columns slightly.

changes are localised chiefly to the parts supplied by the anterior arterial system of the spinal cord. Fig. 25 indicates the arteries of the cord and their branches, which are seen on transverse section. Fig. 27 shows the regions supplied by the anterior and by the posterior system of arteries of the spinal cord respectively. Fig. 26 shows the regions supplied by the central and by the peripheral arteries of the cord.

In acute anterior poliomyelitis, in its most limited form, the changes are often located to the region supplied by the central system of spinal arteries, frequently around the anterior central artery of the cord in the grey matter of the anterior horn. In other cases, as Marie points out, they are found chiefly around the anterior root arteries as they enter the anterior horns of grey matter (*see* Fig. 28). These arteries of the cord are end-arteries. The anterior median artery does *not* usually divide at the bottom of the anterior fissure, but passes to one anterior horn only. Hence, if the lesion were chiefly in the distribution of one anterior median artery, only one horn of grey matter would suffer, i.e. the one to which this artery passed.

Sometimes the pathological changes extend to the white matter just around the anterior horn; this is explained by the fact that the branches of the anterior central artery are not all limited to the grey matter, since terminal branches pass to the white matter just around the anterior horn (*see* Fig. 26).

Often the changes are limited to one portion of the anterior horn (*see* Fig. 28), whilst at another level a different part of the horn may be affected. In old cases, as already mentioned, the degeneration in the nerve cells follows the course of vessels in the anterior horn, and is not localised to definite cell groups. The relation of the disease to the vascular supply is also shown in early cases, by the cell infiltration in the walls of the vessels, and around the vessels, in the anterior median fissure of the cord, *before* the anterior median artery enters the cord substance. As Schultze points out, acute anterior poliomyelitis appears to be a special type of myelitis localised to the region of distribution of the anterior system of spinal arteries, and especially to the anterior central artery and its branches. This region of the cord (anterior grey matter) is the part which has the least firm structure. It is also the most vascular part of the transverse area of the cord. Further, the anterior median and its central branches are the largest arteries entering the cord, and as already mentioned they are terminal arteries. Lamy has found that by the injection of indifferent powders into the aorta, embolism of small arteries, followed by hæmorrhage and softening, can be produced in the grey substance of the cord. From the sudden onset of the paralysis, Marie has suggested that an infectious embolism or thrombosis in one of the branches of the anterior median artery, or in one of the anterior root arteries, may be the starting point of the disease. The thrombosis detected by Batten has been already mentioned. In a case already referred to, which I examined most carefully, I was unable to find any trace of embolism, thrombosis, or hæmorrhage in the anterior horns, though the onset of the paralysis was exceedingly sudden, and appeared to point to arterial obstruction. In many cases recently examined no thrombosis or hæmorrhage could be detected. From a careful study of the pathology Wichman concludes that the disease is an infiltrative myelitis; interstitial changes are the most marked; parenchymatous changes (degeneration of ganglion cells) without interstitial changes were not observed by him.

Goldscheider has suggested that in acute anterior poliomyelitis substances giving rise to cell proliferation affect the walls of the blood vessels by filtration and diffusion from the blood. In the early pathological changes the infiltration of the adventitial and perivascular sheaths is the most prominent feature.

Woodhead, in his interesting address at the meeting of the British Medical Association, 1894, drew attention to the importance of the cell walls of the vessels in pathology. According to Heidenhain the cells of the capillary walls act as true secreting cells. When stimulated there

is an increased flow of lymph into the surrounding tissue, and whenever this is greatly increased there is also a wandering out of leucocytes, or, at least, an accumulation of leucocytes near the walls of the vessel. Certain substances appear to have the power of stimulating the endothelial cells of blood vessels. The toxins produced by certain micro-organisms probably have this power.

Woodhead points out that the endothelial walls of the cerebro-spinal system of capillaries and lymphatics are extremely delicate and active; the lymph flow is great; and if the products of bacteria or bacteria themselves have any effects on the walls of capillary vessels they should make themselves manifest here, and we should expect to find well-marked vascular and peri-vascular interstitial changes. Owing to the greater vascular supply a toxin in the blood will be brought into closer relationship with the grey matter than the white.

The peculiarities of the lymphatics of the cord may also be of some importance in the production of the pathological changes—there are no true lymphatic vessels, but a lymph space ensheaths the spinal vessels.

A few years ago Schultze detected the Jäger-Weichselbaum's diplococcus in the cerebro-spinal fluid obtained by lumbar puncture in an early case of infantile paralysis; and in eight cases, recorded subsequently by other observers, diplococci or forms of the meningococcus have been found in the cerebro-spinal fluid; but it has not yet been definitely proved that any one of the organisms found has been the cause of the disease.

Experiments on animals, made during recent years, have shown that various forms of myelitis can be produced by the injection of bacteria and of toxins formed by bacteria.

It is probable that the toxins produced by the action of bacteria may cause myelitis in man, since toxins free from bacteria can produce myelitis in animals. Also bacteria cannot usually be detected in the cord when myelitis has been produced experimentally by the injection of micro-organisms. The toxin appears to act in some cases chiefly on the vessel walls, in other cases directly on the nerve parenchyma, or on both structures. Marinesco has shown that in myelitis produced by bacteria the micro-organisms disappear from the spinal cord after a few days. In poliomyelitis the cord has been examined by Marinesco and others for micro-organisms with negative results.

Marinesco produced poliomyelitis in animals by injecting micro-organisms into the blood vessels which directly supply the spinal cord. He has also produced changes like those of acute anterior poliomyelitis by the injection of streptococci and of the influenza bacillus within the spinal dura mater. A number of observers have produced spinal changes in animals resembling those of acute poliomyelitis by the injection of various bacteria (typhoid, influenza, diphtheria bacilli, erysipelas streptococci, and the bacterium coli).

It appears probable, that acute anterior poliomyelitis is due to the

action of a toxin circulating in the blood, which is produced by micro-organisms. The infective agent or organism may find its way into the system through the gastro-intestinal tract, since the disease occurs most frequently at a time of the year when gastro-intestinal affections are most common in children. There is also the possibility of organisms gaining entrance into the system through the nasal mucous membrane or the middle ear.

The **etiology** of acute anterior poliomyelitis also throws some light on its pathogenesis.

The two sexes are affected with about the same frequency, and there is no hereditary tendency to the disease.

Acute anterior poliomyelitis may occur at any age, but in the majority of cases the onset is during the first three years of life (472 out of 609 cases collected by Allen Starr). The onset is most frequent in the second year, after the fourth year it is much less frequent. In the adult it is very rare: amongst 83 cases of acute anterior poliomyelitis Schultze met with 2 cases only in the adult. The disease occurs most frequently during the hot season of the year, as pointed out long ago by W. H. Barlow, of Manchester. Amongst 452 cases collected by Allen Starr, the onset in 49 cases was in June, in 97 in July, in 116 in August, in 65 in September. Thus, in 327 out of the 452 cases the onset occurred between June and September.

Exposure to cold has sometimes appeared to be an exciting cause. Starr has seen several cases in boys following swimming in cold water for a long period. The writer has met with two cases presenting this history.

Occasionally the disease has followed a fall; this may be the exciting cause in rare instances.

Sometimes it has directly followed an acute infectious disease, such as measles, scarlet fever, whooping cough, diphtheria, etc., and probably the infectious ailment has played some part in the causation of the paralysis.

As already mentioned, there are often gastro-intestinal symptoms at the onset, before the paralysis is noted. I have met with a case in which these were very marked and prolonged for two weeks.

During the last fifteen years a number of small epidemics of acute anterior poliomyelitis have been recorded, in which many cases of the disease, 13, 30, 45, 54, etc., up to 126 have been observed in one locality in the comparatively short period of two or three months. (These epidemics are recorded in the writer's article in the *Practitioner*, May 1902, and in the article by E. F. Buzzard, *Brain*, 1907.)

Acute Anterior Poliomyelitis of the Adult.—This is a rare affection. Probably many of the cases, in which this diagnosis has been made clinically, have been really due to multiple neuritis. But in a few cases the clinical diagnosis has been confirmed by pathological examination.

The age of the patient is usually from twenty to thirty. At the

onset of the affection febrile symptoms are usually present. As the fever diminishes paralytic symptoms develop acutely. Usually the paralysis is much more extensive than in the infantile form. All four limbs are often affected ; in other cases both legs only. The bladder and rectum are not affected ; bed-sores do not develop. The paralysed muscles undergo atrophy rapidly, and show the same electrical changes as in infantile paralysis. There is no anæsthesia, but there may be slight pain and paræsthesia in the limbs at the onset. The knee-jerks are lost when the legs are paralysed. Complete recovery is very rare.

In a case which I have recorded the symptoms commenced by " numbness " in the right hand. This extended to all the limbs. Then the right arm became paralysed, afterwards the left, and finally all four limbs were completely paralysed. The knee-jerks were absent ; there was no anæsthesia ; the bladder and rectum were unaffected. Death occurred in five weeks.

In only a very few cases has the diagnosis of acute anterior poliomyelitis of the adult been confirmed pathologically. In the case just mentioned, I found the following pathological changes :—

In the anterior horns of the grey matter of the spinal cord on both sides, in cervical, dorsal, and lumbo-sacral regions, changes were present; but they were slight in the dorsal region. There was great dilatation of the blood vessels, their sheaths were distended with round cells, and there was well marked cell infiltration (leucocytes and compound granular cells) around the vessels in the lumbar, sacral, and cervical regions. The exact extent of the changes in the anterior horns varied slightly at different levels, but the chief seat of the cell infiltration was the outer part of the anterior horns (*see* Fig. 99).

The changes were those of an interstitial inflammation, and followed the course of the blood vessels ; they were found in the area of distribution of the *anterior spinal* system of arteries, more or less, throughout the whole length of the cord. In the lumbar, sacral, and cervical regions there was an area of myelitis at the outer part of each anterior horn. In this area the cell infiltration was marked and nerve fibres and cells were destroyed.

In many sections of the *dorsal* region of the cord the vascular changes were marked, whilst other changes were slight. The vessels of the anterior horns were dilated, and at the outer half of each anterior horn the peri-vascular sheaths were distended with round cells. The nerve cells of the outer half of each anterior horn only presented the slightest changes, and there were only a few scattered round cells just around the vessels.

In all regions the vascular changes were in great excess of the other myelitic changes. In many sections, the vessels of the anterior and posterior commissures and of the inner part of the anterior horns were also greatly dilated, and their sheaths distended with round cells, whilst the cell infiltration was limited to the outer part of the anterior horns.

The intensity and wide distribution of the vascular changes were strongly suggestive of a vascular origin of the disease, and the remarks made already, with reference to the irritation of endothelial cells of small vessels, would apply to the case of acute poliomyelitis in the adult.

A few similar cases have been recorded (*see* References).

It appears probable that the pathology of the disease is the same as that of infantile paralysis and that the cause of the changes is some micro-organism or the toxin produced by a micro-organism. The changes in the cord appear to be primarily inflammatory and the degeneration of nerve cells secondary.

Diagnosis.—The chief difficulty is in distinguishing these cases from peripheral neuritis. The absence of anæsthesia and of muscular hyper-

FIG. 99.—Section of spinal cord stained with logwood. Lumbar region showing acute anterior poliomyelitis of the adult. Outer part of anterior horn affected. *a a*=dilated blood vessels; the lymph sheath distended with round cells.

æsthesia are in favour of poliomyelitis. Pains in the limbs are often absent in poliomyelitis and when present are not severe, whilst they are usually severe in peripheral neuritis. Also the development of the disease is more gradual in peripheral neuritis.

In acute myelitis there is usually affection of sensation, and of the bladder and rectum, and bed-sores often develop: all these symptoms are absent in poliomyelitis.

In spinal hæmorrhage the paralysis may closely simulate acute anterior poliomyelitis, but the onset of hæmorrhage is more sudden, there are no febrile symptoms, sensation is usually affected, especially the sensation for pain and temperature, often the bladder and rectum

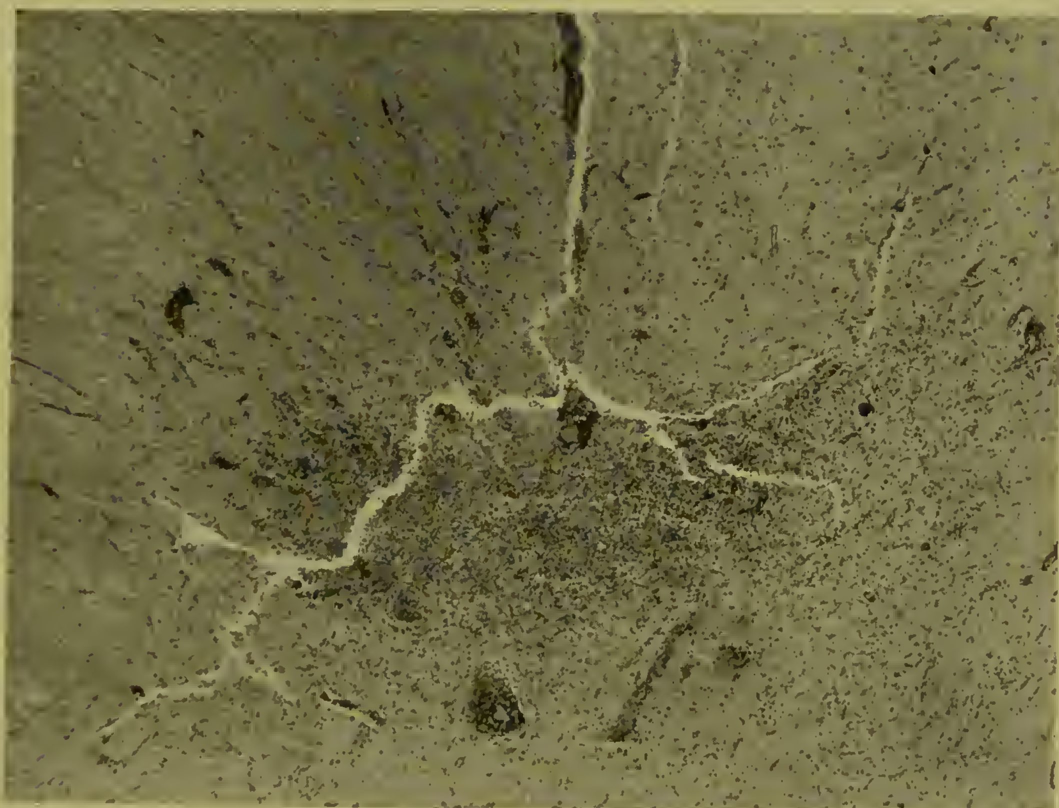


Fig. 100.—Microphotograph of Anterior Horn of Grey Matter of Spinal Cord, in acute anterior poliomyelitis of the adult. Logwood stain. Grey matter at lower part of section: white matter at upper part. Note cell infiltration (marked with black dots) at outer part of grey matter, about middle of photograph. Note cell infiltration around blood vessels, well seen around transverse section of a blood vessel at lower part of photograph.

are paralysed, the knee-jerks are often increased and ankle-clonus and the extensor type of plantar reflex often develop.

Loss of the vibrating sensation would be evidence against acute anterior poliomyelitis and in favour of peripheral neuritis or myelitis.

Prognosis.—The disease very rarely causes death, and in such cases the fatal termination is usually due to paralysis of the respiratory muscles. Complete recovery is very rare, though considerable improvement in the paralysed limbs and gradually restriction of the paralysis is common.

Treatment.—I. At the *onset* of acute anterior poliomyelitis, whether of

the infant or of the adult, the patient should be placed under those conditions which appear to be most likely to prevent further extension of the changes in the spinal cord. He should be kept at rest in bed, and it is more advisable for him to be kept on the side, or in the prone position, than on the back. It is also important that the bowels should be kept quite regular by some mild purgative.

Diaphoretic treatment is recommended by many writers, and appears to be justifiable. The skin should be made to act freely by well surrounding the body with woollen clothing, blankets, etc., by the use of warm bottles, and by giving hot fluids. But it is not advisable to carry the patient to a warm bath.

Sodium salicylate is recommended during the febrile stage, and it is possible that it may be of service, though very definite evidence in its favour has not been produced.

II. The patient very often comes first under treatment *after the acute stage has passed.* Then the question arises, Can anything be done to restore the function of the paralysed muscles ? No drug is known to be of definite service, though strychnine has been often given by mouth and hypodermically. Some physicians also assert that electrical treatment and massage are useless, or almost useless. This is a most convenient view to hold ; it saves the parents, the medical man, and the patient a great amount of trouble and time. But to the writer it does not appear to be a fair view to take.

As regards *electrical treatment*, it is of course difficult to bring forward definite proofs of its value. There can be no doubt that in the majority of cases it is not capable of producing *great* improvement *rapidly.* Also there is no evidence that electricity has any influence on the spinal cord changes. Nevertheless, it appears probable that electrical treatment of the paralysed muscles is of service ; and it is only fair to the patient to give a satisfactory trial to this method of treatment. There are rare cases on record in which electrical treatment has been followed by considerable improvement, even when it has been commenced many months or several years after the onset of the paralysis. The nerve cells supplying a muscle of the limb are usually rather widely distributed in the cord in the vertical direction and often extend through two or three spinal segments. Hence in a small localised poliomyelitis often many of the cells sending fibres to a paralysed muscle escape destruction. Some of the fibres of the muscle do not degenerate and the function of these can be improved by electrical treatment. By electrical treatment and massage of the paralysed muscles, something may be done to keep up their tone and to prevent or check atrophy from simple disuse, so that if the nerve cells in the anterior horns should ever regain their functions, the muscles which they supply will not be too wasted to take on their actions. Whenever electrical treatment can be carried out, it should be commenced after the third week, and continued for a long period. At first, for a few days, it is best to apply the electrode sponges to the limb

with the current broken, so that the child may become accustomed to the sight of the battery and sponges before they cause any pain. If the current is allowed to pass through the limb at the first application, the child is frightened, and will probably scream and struggle afterwards whenever he sees the battery. Galvanism is the form of electricity commonly used, and it is especially suitable for paralysed muscles which give the reaction of degeneration, and therefore do not contract to faradism. These muscles react better to the anode pole, therefore the kathode electrode should be placed on the back, or breast, or other indifferent part, and the anode electrode over the region of the paralysed muscles. The anode sponge may be rubbed over the muscles, or it may be placed on the muscles and the circuit made and broken at very short intervals. It is important that the strength of the current should be tested by placing the sponges on the hand before they are applied to the patient. Strong currents should be avoided ; a current which is just sufficient to cause a contraction of the muscles is strong enough. Usually a current of two to five milliampères is sufficient. It should be applied once or twice daily for a few minutes to each group of paralysed muscles. Large electrodes are advisable.

For those muscles in which the faradic excitability has not been lost the faradic current has been occasionally employed.

The use of the electric bath for the limb is strongly recommended by L. Jones. The current from a galvanic battery or the electric light current (reduced in strength) may be employed for the bath.

A second method of keeping up the nutrition of the paralysed muscle is by massage, which improves the circulation of blood and lymph in the affected limb. The limb should be rubbed and stroked in an upward direction (i.e. towards the trunk), passive movements should also be made, and afterwards kneading and pinching of the muscles. Rubbing of the limb with some liniment containing spirit aids the circulation and makes the skin warmer.

Unless the parents are sufficiently wealthy to engage a trained nurse, the mother should be taught to carry out the electrical treatment and massage herself. The important point is perseverance in the treatment for a long period. If there is no improvement in the muscles after electricity and massage have been carefully employed for one year, it is useless to continue the treatment longer.

The muscles in which there is some return of power should be used frequently, and systematic gymnastic exercises should be carried out so as to increase their development, and to enable them, to some extent, to supplement by their action the adjacent paralysed muscles.

The paralysed limb should be kept warm by means of a thick woollen stocking or by surrounding it with cotton wool. The use of a warm bottle at night is desirable in cold weather, and warm salt baths for the limb are also of service. The position of the limb should be attended to, so as to check the development of deformities. If the leg be paralysed,

the child should not be allowed to sleep or lie with the knees and hips flexed. The foot should also be protected by a cradle from the weight of the bed-clothes when the anterior tibial muscles are paralysed, so as to check the development of talipes equinus or equino varus. An attempt should be made to prevent the foot dropping, and to keep it at a right angle to the leg. With this object, a small elastic band (artificial muscle) is attached at one end to a leather strap or bandage encircling the anterior part of the foot, and at the other end to similar leather straps encircling the leg and the thigh (Barwell and Wright). Also "Scarpa's shoe" may be of service in certain cases.

III. When the paralysis has been present for some years, and there is no hope of further improvement, the question of orthopædic surgery should be considered. Various mechanical apparatus are employed for giving support to loose joints, and special boots are employed for limbs that are too short.

Tenotomy is often of much service in removing deformities due to over-action of the opponents of paralysed muscles. Thus, division of the tendo Achillis may be required on account of talipes equinus, and various tendons may be divided with advantage in other deformities. In certain cases, when a joint is very lax, the production of anchylosis of the affected joint may enable the patient to get about with less difficulty. Thus, by causing fixation (anchylosis) of a lax and flail-like ankle joint the patient is able to walk with less difficulty. The operation (arthrodesis) should, of course, only be performed by a surgeon who has devoted special attention to this branch of surgery.

During the last fifteen years the operation of *tendon transplanting* or *grafting* has been employed with great success in suitable cases. This operation consists in attaching the tendon, or a portion of the tendon, of a normal muscle to the tendon of an adjacent paralysed muscle. The normal muscle on contracting then produces traction on the tendon of the adjacent paralysed muscle. There are many ways of performing these operations. The paralysed muscle may be divided and its tendon fixed to that of an adjacent normal muscle; but a more successful method has been to divide the tendon of the normal muscle completely or partially, and attach it, or a slip of it, to that of the paralysed muscle.

The operation was first performed and recommended by Nicoladoni, and many years afterwards its value was also demonstrated by Drobnik. More recently many cases have been recorded by Vulpius, Robert Jones, Eve, Lange, Tubby, and others.

In talipes equino varus, due to paralysis of the extensor longus digitorum and peronei, the tendon of the normal extensor longus hallucis has been divided and attached to the adjacent tendon of the paralysed extensor longus digitorum. In other cases the tendon of the normal sartorius has been inserted into the tendon of a paralysed quadriceps, and various other active muscles of the leg and arm have been attached to the tendons of adjacent paralysed muscles.

In talipes equinus (from infantile paralysis) the tendo Achillis has been split longitudinally. The outer half of the tendon has been cut across just above the ankle, and attached to the paralysed peroneus longus, which had been also cut across (Tubby). In paralysis of the quadriceps, the tendons of the flexors of the knee (biceps, semi-membranosus and semi-tendinosus) have been divided, and attached to the tendon of the quadriceps just above the patella (Krause).

When the action of the normal muscle is closely allied to that of the paralysed muscle to which it is attached, the patient is soon able to accomplish voluntarily the movement of the latter. Eve points out that the graft tendon also acts mechanically as a supporting band applied to the ankle or foot, or as a support to the lax tendon of a paralysed muscle. When one or more of the contracted antagonists is chosen to reinforce the paralysed muscle, the antagonist is weakened as if tenotomy had been performed, and thus the operation acts favourably in two opposite directions (Eve).

In the method of Goldthwaite a longitudinal incision is made into the tendon of the paralysed muscle, and into this incision is inserted and fixed the cut end of the tendon of the normal muscle.

Lange sutures the divided tendon of an active muscle, not to a paralysed tendon, but directly to the periosteum of a suitable bone. Thus, when the extensor longus digitorum is paralysed, but the tibialis anticus is active, the tendon of the latter is attached to the periosteum of the dorsal surface of the cuboid bone. The most important action of the extensor longus digitorum is dorsiflexion of the foot, and by the new attachment of the tibialis anticus this action can be performed. Lange claims that by this method there is not the risk of the stretching of the tendon of the paralysed muscle. Others also prefer this operation.

Drobnik attaches great importance to massage of the muscles after the operation. It improves the nutrition of the limb, and prevents adhesion with the skin.

R. Jones points out that the action of antagonistic groups of muscles is not the sole cause of paralytic deformities. Several factors are instrumental in producing them, such as gravity, body weight, shape of articular facets, and to a less degree unbalanced muscular action. Before attempting to treat a weakened set of muscles he recommends that we should prevent them from being overstretched, and maintain them in a slackened posture, by extension at the knee, and by rectangular fixation at the ankle.

For the exact methods of performing the various operations of "tendon grafting" recent surgical literature must be consulted. These operations should only be performed when there is no hope of further improvement by medical treatment. In suitable cases, however, they have proved to be of great service.

The treatment of acute anterior poliomyelitis of the adult is the same as that of the disease in the infant. F. Muller recommends the injection of ergotin and atropine.

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CHRONIC AND SUBACUTE ANTERIOR POLIOMYELITIS

This very rare disease differs clinically from acute anterior poliomyelitis simply in its gradual onset. Many of the cases recorded years ago were probably due to multiple peripheral neuritis; but there are on record a few cases in which the diagnosis of chronic poliomyelitis has been verified pathologically.

Etiology.—The patients have usually been adults. The cause of the disease is unknown. But probably in some cases the disease is due to the action of a toxic substance on the nerve cells of the anterior horns. Degeneration of these cells has been caused by chronic lead poisoning in experiments on animals; also in man chronic poisoning by lead and by arsenic have sometimes caused degeneration of the nerve cells of the anterior grey matter, as well as degenerative neuritis in the peripheral nerves. (Nonne has reported one case in a diabetic patient.) Possibly injury has been the starting point of the disease in rare cases, by causing minute hæmorrhages or vascular changes in the anterior grey matter. Cases have been attributed to over-strain and to syphilis. In some cases chronic anterior poliomyelitis has developed in patients who have suffered from acute anterior poliomyelitis many years previously (*see* p. 201).

Symptoms.—The disease commences with weakness in one limb: in course of time—weeks or months—another limb is affected in the same manner. In some cases both legs are affected first; in other cases one arm is first affected, then the other arm, or the leg on the opposite or on the same side. But in the course of months or years all four limbs are affected. The muscles of the legs (below the knee) or the upper arm muscles are first affected.

The loss of power gradually increases until the affected muscles are

completely paralysed. The paralysed parts are flaccid. The muscles undergo atrophy, and present, on electrical examination, the reaction of degeneration. The knee-jerks disappear, and the other reflexes are diminished or lost. Fibrillary twitchings are seen in the muscles affected. There is no anæsthesia and no sensory symptoms occur, except slight "rheumatic pains." There is no pain on pressure of the muscles.

The bladder and rectum are not affected, and bed-sores do not develop. Sometimes there is diminution of perspiration (Strümpell).

In rare cases the paralysis has extended to the muscles of the neck, lips and tongue, and the muscles of deglutition.

An important feature of chronic poliomyelitis is the fact that paralysis of muscles occurs *before* atrophy. (In progressive muscular atrophy the wasting is the primary symptom and loss of power follows, and is the result of the wasting.) It is said, however, that transitional cases sometimes occur.

The disease is usually progressive. The symptoms reach their complete development in from one to four years or longer. The intercostals and diaphragm become finally paralysed, and death occurs from respiratory paralysis. Sometimes death has been due to intercurrent affections (tuberculosis, pneumonia, bronchitis).

The disease is stated to become arrested occasionally, and improvement or recovery has been reported. It is at present impossible to say whether such cases have been poliomyelitis or peripheral multiple neuritis.

The **prognosis** is worse the more gradual the development of the symptoms. When the respiratory muscles become affected there is great danger of death from asphyxia. When the muscles of deglutition are involved there is danger of broncho-pneumonia occurring through food finding its way into the air passages.

Diagnosis is based on the gradual development of motor paralysis, which is *followed* by muscular atrophy. The muscles affected present the reaction of degeneration on electrical examination. There is no affection of bladder or rectum, no form of anæsthesia, and the sensory symptoms of peripheral neuritis are absent.

In progressive muscular atrophy the symptoms are also entirely motor, and the lesion is also in the anterior horns of grey matter. But in progressive muscular atrophy wasting of the muscles is the first symptom, and loss of power, and finally paralysis, is the *result* of the muscular atrophy. (As just stated in chronic anterior poliomyelitis loss of power or paralysis occurs before the atrophy.) In progressive muscular atrophy the wasting usually begins in the small muscles of the hand; it spreads very slowly from muscle to muscle. In chronic anterior poliomyelitis the leg muscles or upper arm muscles are first affected; several muscles or a portion of a limb are affected together, and the course is not so slow as in progressive muscular atrophy.

In amyotrophic lateral sclerosis the symptoms are entirely motor,

but the legs present the symptoms of spastic paralysis—the knee-jerks are increased, ankle-clonus and Babinski's extensor type of plantar reflex are present, and there is rigidity to passive movements; also the atrophic paralysis affects first the small muscles of the hand.

In syringomyelia the presence of sensory symptoms distinguishes the case from chronic anterior poliomyelitis. Also in many cases of syringomyelia the legs are spastic.

The diagnosis from peripheral neuritis is made by the presence of sensory symptoms in the latter disease, i.e. hyperæsthesia, well marked pains, numbness, paræsthesia, or anæsthesia, tenderness and pain on pressure over the muscles and nerve trunks. All of these symptoms are absent in chronic anterior poliomyelitis. In the later affection

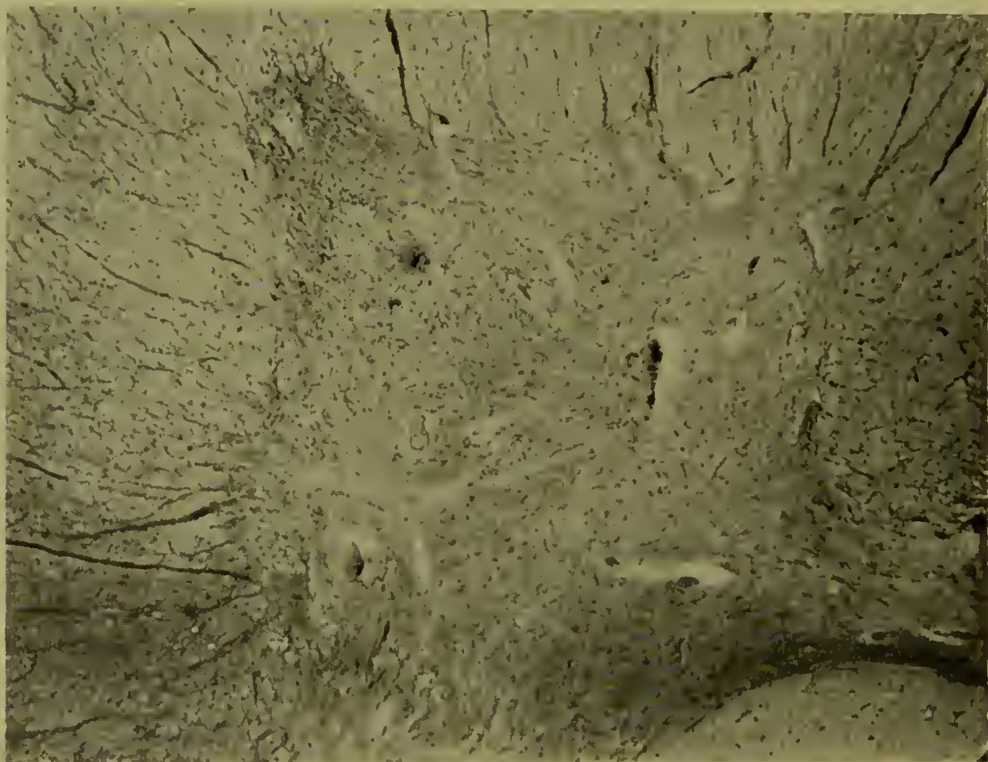

FIG. 101.—Chronic Anterior Poliomyelitis. Microphotograph of anterior horn of grey matter; carmine stain; showing absence of nerve cells. Compare with anterior horn of normal cord (Fig. 113, p. 244).

there is no history of the usual causes of peripheral neuritis, and there are none of the mental symptoms so common in alcoholic neuritis.

Pathological Anatomy.—The diagnosis of chronic or subacute poliomyelitis has been verified pathologically, but only in a few cases (Dreschfeld, Oppenheim, J. B. Charcot and Dutil, Nonne, Strümpell, Bielschowsky). In these cases examination of the spinal cord has revealed atrophy and degeneration of the nerve cells and fine nerve fibres in the anterior horns of grey matter. The white substance is normal, or shows only scattered atrophied fibres in the neighbourhood of the anterior grey substance. The nerve cells of the posterior horns and of Clarke's columns show scarcely any change. The anterior nerve roots are de-

generated. The paralysed muscles are atrophied and degenerated. The transverse striation is often lost. The muscle sheath, in advanced cases, may be almost empty and contain only fragments of degenerated fibres. The inter-muscular connective tissue and fat may be increased. Usually only very slight changes are found in the peripheral nerves (secondary degeneration).

In old cases the neuroglia and vessels in the anterior horns show little change, but in more recent cases both show inflammatory changes. In the case recorded by Dr. Dreschfeld (which I had the opportunity of observing at intervals for several years) there were marked vascular changes; dilatation of blood vessels and thickening of their walls, in addition to atrophy of ganglion cells in the anterior horns. In the case recorded by Dutil and Charcot there was also marked thickening of the walls of the anterior spinal and anterior root arteries.

[In the case recorded by Bielschowsky the patient had suffered for nine years from atrophic paralysis of gradual development, affecting the muscles of the limbs and trunk. The leg muscles were first affected, and the disease extended upwards until the muscles of the neck were implicated, and the tongue slightly. Paralysis preceded atrophy, and the former was always more marked than would correspond to the degree of atrophy.

The pathological examination showed changes in the motor ganglion cells of the anterior horns of the spinal cord at all regions. The intensity of the changes varied widely in the same cell group. The changes were most marked in the parts supplied by the central artery. At the periphery of the grey matter of the anterior horn, which receives small vessels from the peripheral arterial system of the spinal cord (*see* Fig. 28) cells were found at all regions which presented a normal or almost normal appearance. At the affected parts of the anterior horn, where the changes were most marked, fine nerve fibres, cells and all structures were degenerated. Bielschowsky attaches great importance to the relation of the changes to the blood vessels.]

It is probable that in some cases the changes are *degenerative*. They affect chiefly the motor ganglion cells of the anterior horn which undergo simple atrophy, whilst the blood vessels appear normal. In other cases, *interstitial inflammatory form*, there is a chronic or subacute myelitis (allied in nature to acute anterior poliomyelitis) limited to the anterior horns of grey matter. In this latter group of cases, at the early stage the vessels of the anterior horn are dilated and full of blood, especially the branches of the anterior median artery. The vessel walls are thickened, sometimes hyaline, and their nuclei increased. The nerve cells are diminished in size; their processes have disappeared, and the Nissl's granules, especially at the periphery of the cell, present a "dusty" appearance. In the interstitial tissue spider tissue cells are seen. At the advanced stage the vessels are thickened and sometimes hyaline; they are no longer so distended with

blood; the neuroglia tissue is increased, its nuclei are more numerous, and many spider cells are seen. The nerve cells are atrophied and often entirely absent; those remaining are small, rounded and without processes; the Nissl's granules are indistinct.

In a case reported by Dejerine and Sottas there were changes both in the peripheral nerves and in the nerve cells of the anterior horns; but the latter were much less marked than the former, and the case is regarded as one of motor peripheral neuritis with consecutive spinal lesion. Schuster reports a similar case in which the peripheral nerves, the anterior horns, posterior roots and Burdach's columns were affected. (See description of toxic degeneration of lower neurons.)

Treatment.—We know of no means of arresting or curing this disease. At the early stage overstrain should be avoided: rest is advisable; strychnine, galvanism and warm baths have been recommended, but there is no clear evidence of their value. Hypodermic injection of strychnine is worthy of fair trial in all cases (*see* p. 249).

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TOXIC DEGENERATION OF THE LOWER NEURONS

S. Barnes has furnished clinical and pathological evidence of a type of paralysis which resembles multiple neuritis, but is associated with great atrophy of the hand muscles. The disease begins usually about the second or third week after a febrile affection; sensory symptoms are present, but slight in degree; the condition resembles somewhat progressive muscular atrophy, but the etiology is different, and there is a "constant tendency to improvement." Barnes thinks the condition is one of toxic degeneration of the lower neurons, especially the motor neurons. The pathological examination of one case which he records supports this view. There was degeneration both in the peripheral nerves of the arms and in the nerve cells of the anterior horns of grey matter. The changes in the nerve cells were most marked in the anterior horns of the lower cervical and first dorsal segments: some of the cells had atrophied, others were in a condition of tigrolysis. In the lumbar region there was tigrolysis, but no atrophy of nerve cells.

Clinically the cases differed from multiple neuritis, in its usual form, by the great atrophy of the small muscles of the hands and by the comparatively slight sensory changes.

I have recorded a similar case, in which paralysis and atrophy of the small muscles of the hand and of the extensors of the fingers and wrist, with paresis of the flexors, followed an acute illness (? influenza). The hand muscles were most affected. There was no evidence of lead poisoning, or of other common forms of peripheral neuritis. Recovery occurred. Since the case was recorded (in 1903) there have been two similar attacks, in which the muscles of the hand and forearm were affected, and one attack in which the forearm muscles only were involved. Complete recovery has followed each attack.

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LANDRY'S PARALYSIS: ACUTE ASCENDING PARALYSIS [1]

Many years ago Landry described a peculiar form of paralysis, which began in the legs and rapidly extended to the trunk and arms: there was no atrophy of muscles, no affection of bladder and rectum, and no affection of sensation: on pathological examination no changes could be detected in the nervous system. During the last forty years many cases of acute paralysis have been recorded as Landry's paralysis, which were probably due to acute anterior poliomyelitis, peripheral neuritis, or other lesions.

With two exceptions, all the cases, supposed to be due to Landry's paralysis, which have come under the writer's observation, have been clearly shown, by clinical or pathological examination, to be due to some other disease.

Most of the cases diagnosed as Landry's paralysis are found to be due to multiple neuritis or acute anterior poliomyelitis on careful pathological examination. The question arises, are all cases diagnosed as Landry's paralysis due to these diseases?

Certainly the greatest caution should be observed before giving a diagnosis of Landry's paralysis. But recent careful pathological and clinical records appear to show that there is a disease to which the name of Landry's paralysis may be conveniently applied. The symptoms correspond to those of Landry's description, and though pathological examination reveals slight changes in the cord, they can only be detected by the delicate methods of Nissl and Marchi, and the peripheral nerves do not present the changes of neuritis. The disease has occurred most commonly in adult males. It has sometimes followed exposure to cold and wet, or infectious fevers.

The **Symptoms** usually commence with paresis in the legs, and there is often pain in the back and numbness or tingling in the limbs. The

[1] Although Landry's paralysis does not cause muscular atrophy, the description of the disease has been inserted here because the symptoms somewhat resemble those of acute anterior poliomyelitis of the adult.

legs rapidly become paralysed, then the trunk muscles, and afterwards the muscles of the arms and neck, and the muscles of respiration, deglutition and articulation become paralysed. The deep and superficial reflexes are lost, the limbs are flaccid, but the muscles do not atrophy, and there is no reaction of degeneration. There is no anæsthesia, and the sphincters are not affected. Bed-sores do not occur. Death usually occurs in a few days (two to fourteen).

Febrile symptoms and slight enlargement of the spleen often occur. In rare cases the paralysis has a descending course.

Pathology.—In recent years, a number of most careful pathological examinations have been made.

No changes can be detected in the nervous system by the naked eye, except general vascular engorgement. In the peripheral nerves there are no neuritic changes. In the spinal cord the only changes are in the nerve cells of the anterior horns and of Clarke's columns. These changes have usually been slight, and detected only by Nissl's method of staining. E. F. Buzzard has recorded early pericentral chromatolysis in a considerable number of the cells in the regions mentioned; and in a few loss of chromatin granules and eccentration of the nucleus, especially in the lumbar region. Chromatolysis of the nerve cells of the anterior horn has also been recorded by Sheppard, Walker Hall and others. In many cases no changes have been detected, either in the cord or peripheral nerves.

The relation of the disease to various micro-organisms has not yet been definitely decided; but it appears probable that the affection is due to the action of some toxin on the lower motor neurons (*see* Buzzard's paper). It is interesting to note that in myasthenia gravis, and in cerebral tetanus there are also marked paralytic symptoms without decided pathological changes in the motor nuclei of the medulla, or in other parts of the nervous system.

The similarity of the symptoms to those of acute anterior poliomyelitis, and the slight changes found in the nerve cells of the anterior horns, suggest the view that Landry's paralysis is a very acute form of anterior poliomyelitis, due to a toxin so powerful that the lower motor neurons lose their function and death results before cell infiltration occurs.

The chief difficulty in **diagnosis** is to distinguish Landry's paralysis from acute anterior poliomyelitis. E. F. Buzzard points out that the escape of a single muscle or group of muscles in a paralysed region, or a marked asymmetry in the paralysis on the two sides of the body would be in favour of acute poliomyelitis. In acute poliomyelitis within a few days or a week, if death has not occurred, there is usually rapid recovery in some parts and early atrophy and electrical changes in others. In Landry's paralysis, if recovery occurs it is slow and evenly distributed, and marked atrophy of muscles and the reaction of degeneration are usually absent.

Multiple neuritis differs from Landry's paralysis by its sensory symptoms—diminished sensibility and marked tenderness of the muscles, both of which are absent in the latter disease. In multiple neuritis the limbs are more affected than the trunk, and the peripheral parts of the limbs more than the proximal ; of the trunk muscles, the diaphragm is affected chiefly, and of the cranial nerves, if any are affected, it is the facial. In Landry's paralysis the muscles of the trunk and limbs are generally and evenly affected, at first the legs only, then the trunk ; the intercostals are affected before the diaphragm ; and when the cranial nerves are affected the muscles of deglutition, phonation and articulation suffer first. There is no localised atrophy and no reaction of degeneration in Landry's paralysis, whilst both are usual in peripheral neuritis (*see* E. F. Buzzard's article).

Treatment is of little service. The drugs which have been recommended are aspirin, sodium salicylate and ergotin. Antisyphilitic treatment is indicated when there is a history of syphilis. Artificial respiration will be necessary if the respiratory muscles become paralysed.

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PROGRESSIVE MUSCULAR ATROPHY

This is an exceedingly rare disease. Many of the cases which would now be regarded as syringomyelia, idiopathic muscular atrophy, and amyotrophic lateral sclerosis were formerly described as progressive muscular atrophy. With the separation of the numerous forms of chronic muscular atrophy the group of cases of progressive muscular atrophy has gradually become smaller and smaller, and some writers regard this affection and amyotrophic lateral sclerosis as varieties of the same disease and would apply the latter name to both.

The chief features of the disease described as amyotrophic lateral sclerosis are atrophic paralysis of the arms with spastic paralysis of the legs. Pathologically the changes may be briefly stated as atrophy and degeneration of the nerve cells of the anterior horns of grey matter in the cervical region, with sclerosis of the crossed or lateral pyramidal tracts. The characteristic features of progressive muscular atrophy are gradual atrophy of the muscles of the arms, with loss of power but without spastic condition of the legs. Pathologically the changes consist mainly in atrophy and degeneration of the nerve cells of the anterior horns of grey matter, chiefly in the cervical region, but without any sclerosis in the crossed or lateral pyramidal tracts.

It must be admitted, however, that in cases presenting symptoms corresponding to those of progressive muscular atrophy (i.e. atrophic

paralysis of the arms without any spastic paresis of the legs) pathological examination has often shown degeneration in the crossed pyramidal tracts. In other words, many cases which clinically present the symptoms of progressive muscular atrophy are found to be cases of amyotrophic lateral sclerosis on pathological examination. So frequently has this been the case, that the two affections are regarded as varieties of one disease by many neurologists, who have always found some change in the crossed pyramidal tracts in the cases of so-called progressive muscular atrophy which they have examined microscopically.

Whilst it must be admitted that the two diseases are closely allied, there are a few rare cases on record which present, both clinically and

Fig. 102.—Claw-shaped Hand. Amyotrophic lateral sclerosis. Note wasting of thenar and hypothenar muscles.

pathologically, the features which have been regarded as characteristic of progressive (spinal) muscular atrophy. To this small group of cases the name progressive muscular atrophy may be fairly applied, and the form in which the small muscles of the hand are first affected is often described as the "*Aran-Duchenne*" type of muscular atrophy.

The **etiology** of progressive muscular atrophy is obscure. Males are more frequently affected than females. The disease begins in middle age; seldom before twenty. It affects mostly individuals whose occupation has been some form of manual labour (i.e. the so-called "working" classes). Overstrain of muscles through heavy work has often preceded the onset of the disease, and has been regarded as a predisposing cause; also it is interesting to note that the disease usually begins in the right arm. Injury, exposure to severe cold, and infectious diseases have been regarded as exciting causes by some authors, and the possibility of a chronic intoxication has been suggested. In favour of the latter view is the fact that chronic poisoning by lead and arsenic sometimes causes atrophic changes in the nerve cells of the anterior horns of grey matter, in addition to degenerative neuritis of

the peripheral nerves. Hereditary tendency to the disease can rarely be traced, though Strümpell and others have met with cases in which two members of the same family have suffered. In very rare cases of infantile paralysis, progressive muscular atrophy has developed at a later period of life (*see* p. 201). Dana thinks that many cases are probably due to syphilis.

The **symptoms** of the disease are chiefly those due to progressive atrophy of muscles. In the majority of cases the disease begins in the arm and usually in the hand. The right hand is generally first affected. The earliest symptom is wasting of the thenar and hypothenar eminences, and of the first interosseous space. These regions become flattened, the eminences gradually disappear, and the first interosseous space becomes more and more depressed (*see* Figs. 102 and 103).


FIG. 103.—Claw-shaped Hand. Amyotrophic lateral sclerosis.

The opponens pollicis, the other thumb muscles (abductor, flexor brevis, adductor), and the first interosseus gradually waste. Loss of power is then noted, but it may be months before this becomes very decided. In course of time the patient is unable to oppose the tip of the thumb and the tips of the fingers, when the latter are kept extended at the phalangeal joints (*see* Fig. 104). Writing and sewing become more and more difficult. The other interossei and the lumbricales become more and more atrophied, and the metacarpal bones are separated by marked depressions.

The interossei of the hand in the normal condition act (1) as adductors and abductors of the fingers ; (2) they cause flexion of the fingers at the metacarpo-phalangeal joints, and extension at the phalangeal joints. When the interossei are paralysed, adduction and abduction of the fingers cannot be performed. At an early stage, however, extension of the fingers causes a little abduction, and may prevent the loss of power of abduction of the fingers being detected. But weakness of the adductors can be detected early. If a pencil be placed between the thumb and first finger, or between two fingers, and the patient told to fix it by adduction (by pressing together the fingers or the thumb and index finger), it is found that when the interossei are paralysed he is not able to retain it in the position indicated, if an attempt be made to withdraw it. The pencil can be easily pulled away from between the fingers, or from between the thumb and first finger ; whilst a healthy

individual can fix the pencil firmly in the position just indicated. This demonstrates the weakness of the adductors of the fingers (interossei, lumbricales). The other action of the interossei is to place the fingers in a position of flexion at the metacarpo-phalangeal joints, whilst they are extended at the phalangeal joints (*see* Fig. 105). When the interossei are much affected the patient is unable to place the fingers in this position (*see* Fig. 106). Also when the interossei are paralysed, over-action of the opponents of these muscles occurs, and the fingers assume a position the opposite of that just described. Therefore there is extension, or over-extension, at the metacarpo-phalangeal joints, and flexion at the phalangeal joints. This produces a claw-shaped

FIG. 104.—Photograph showing, on the left, the hand in amyotrophic lateral sclerosis, when the patient was attempting to oppose the tip of the thumb to the tip of the little finger. To the right is hand of a healthy man when opposing the tip of the thumb and the tip of the little finger.

appearance of the hand ("claw-hand," the *main en griffe* of French writers, and *Klauenhand* or *Krallen-hand* of German writers). Owing to the complete paralysis of the small muscles of the thumb, the over-action of its long extensors often causes the metacarpal bone to be displaced backwards, and to lie in the same plane as the metacarpal bones of the fingers. This is the position of the thumb in the monkey's hand, and in continental works the deformity is described as the "monkey's hand" (German, *Affen-hand*; French, *main de singe*). At a later period the muscles of the forearm atrophy; both the flexors and extensors of the fingers and wrist and the long muscles of the thumb become wasted and paralysed. The movements of the fingers cannot be per-

formed. The claw-shaped deformity of the hand disappears, the fingers lie nearly extended, and the hand is like that of a skeleton (*main de squelette* of French writers). Finally the upper arm muscles become affected.

[The "claw-hand" may occur also in cases of injury to the ulnar nerve, in amyotrophic lateral sclerosis, in syringomyelia and in other diseases affecting the anterior horns of grey matter in the lowest cervical and 1st dorsal segments.]

Fig. 105.—The hand of a healthy man showing action of the interossei—the fingers placed in the position of flexion at the metacarpo-phalangeal joints and extension at the phalangeal joints.

At a late period, after the hand and arm muscles are markedly wasted, the atrophy invades the trapezius (lower $\frac{2}{3}$ first), rhomboids, pectorals, latissmus dorsi and the rest of the scapular muscles. The neck muscles become affected in time, the head is bent forwards and cannot be raised, the chin rests on the upper part of the chest, and the arms hang quite helpless at the side.

In some cases the progress of the wasting is different, and after the hands are affected, the atrophy attacks next the deltoid and shoulder muscles, before the forearm muscles. In other cases the biceps suffer next to the hand muscles.

Fig. 106.—Hand in a case of Amyotrophic Lateral Sclerosis. The patient is attempting to place the fingers in the same position as in Fig. 105. Owing to wasting and paralysis of the interossei the photograph shows that this movement cannot be performed.

In some cases the wasting commences in the shoulder and back muscles first (deltoid, infraspinatus, trapezius and serratus magnus) and gradually extends down the arms.

In very chronic cases the intercostals and diaphragm may be affected finally.

Usually the leg muscles are not affected, or they only suffer at a very late period.

Even when the arms have been quite helpless for years, and the head is bent forwards owing to affection of the neck muscles, the patient may be able to walk quite well. When

the leg muscles suffer those below the knee (usually the peronei) are first affected.

In progressive muscular atrophy the loss of power does not occur before the wasting; the atrophy is followed by weakness, and it is only when the atrophy is complete that total paralysis occurs. The hands are often cold and cyanotic, owing to their hanging position at the side and the complete loss of movement.

Electrical examination reveals a diminution of the excitability both to faradism and galvanism in the affected muscles and in their motor nerves. When the wasting is marked a partial reaction of degeneration is obtained: the jerk of the muscles to galvanic stimulation is sluggish, the ACC is greater or equal to KCC, but the muscles react to faradism. Only in very advanced cases is a true reaction of degeneration obtained, the muscles ceasing to contract to faradism. When the atrophy has become extreme, the reaction to both faradism and galvanism may finally cease.

The reflexes are usually diminished, never increased. The knee-jerks are present; but they may disappear at a late period if the legs become atrophied. There is no ankle-clonus, no rigidity or other sign of spastic paresis of the legs. (Those cases in which there is a gradual development of spastic paresis of the legs with increased reflexes and ankle-clonus, are really cases of amyotrophic lateral sclerosis.) The deep reflexes of the arms are diminished or absent in progressive muscular atrophy, but are always increased in amyotrophic lateral sclerosis (Strümpell).

In the atrophied muscles fibrillary contractions are seen. These consist of slight involuntary twitchings (or rapid contraction) of one or more bundles of the fibres of a muscle, whilst the rest of the muscle is not contracted. The contraction continues for a moment only, then the fibres relax; it is repeated at irregular intervals, it gives rise to a slight flickering of the skin, and various bundles of fibres are affected in succession. The contractions are increased by cold and by mechanical irritation: they are painless and occur in muscles which are not wasted, as well as in those presenting distinct atrophy.

The bladder and rectum are not affected. Sensation is usually unaffected. In a few cases there has been slight pain in the limbs or slight paræsthesia, probably owing to the arms hanging constantly at the side. Anæsthesia of all forms is absent.

In some cases, symptoms of bulbar paralysis have been recorded at a very late period of the disease: also cases have been recorded in which bulbar paralysis has been the first affection and the progressive atrophy of the arm muscles has developed later. But according to Dejerine and Thomas, when bulbar paralysis occurs the disease is amyotrophic lateral sclerosis and not true progressive muscular atrophy.

Course and Prognosis.—Progressive muscular atrophy is a very chronic disease. Often years elapse before the atrophy extends from the hands to the forearms: often there are long periods in which the

disease appears to be almost stationary; but improvement does not occur. The duration is often ten, fifteen or twenty years. Death occurs from some intercurrent disease, such as tuberculosis, acute pneumonia, or broncho-pneumonia; in some cases from asphyxia due to paralysis of the respiratory muscles. (As already mentioned, many cases which at first appear to be progressive muscular atrophy finally present the symptoms of amyotrophic lateral sclerosis.)

Diagnosis.—The characteristic feature of the disease is the progressive wasting of muscles followed by loss of power, owing to loss of muscle substance. The wasting begins in the arms, usually in the small muscles of the hands, rarely in the upper arm muscles. Anæsthesia of all forms is absent; the bladder and rectum are not affected; there is no increase of the knee-jerks; no ankle-clonus; no Babinski reflex; and no spastic condition of the legs.

The diagnosis of progressive muscular atrophy from amyotrophic lateral sclerosis is described on p. 246.

In the clinical group of cases which are described as chronic anterior poliomyelitis, paralysis of muscles (with the reaction of degeneration on electrical examination) occurs before the wasting. Also the course is more rapid. All the muscles of a section of a limb are affected together. The paralysis usually begins in the legs or shoulder muscles first.

In acute anterior poliomyelitis (of the adult or infant) the onset of paralysis is sudden, the small muscles of the hands are rarely affected, the paralysis occurs before the wasting, and usually considerable improvement occurs.

In syringomyelia, the sensory symptoms (analgesia and thermo-anæsthesia) and trophic changes distinguish the disease from progressive muscular atrophy. Also in the former disease, there are often signs of spastic paresis of the legs, and the atrophy is localised to one arm for a longer period.

From lead paralysis the diagnosis is usually easy; but in severe forms of this disease the small muscles of the hand are occasionally atrophied, and in such cases there is degeneration of nerve cells in the anterior horns in the lower cervical region. In favour of lead paralysis would be the blue line on the gums, the history of lead poisoning, and the occurrence of lead colic and other symptoms of plumbism; also the dropped wrist, from paralysis of the extensors, occurs as an early symptom, and the paralysis is greater than can be accounted for by the muscular wasting.

Caries of the cervical vertebræ, cervical myelitis, tumour, or hæmorrhage, may cause atrophic paralysis of the hand muscles. But diminution of sensation, spastic paresis of the legs, and bladder symptoms distinguish the cases from progressive muscular atrophy. Also in cervical caries there is generally prominence of one or more vertebral spines, pain on percussion of the spine at the seat of the disease, and the X ray photograph may show signs of bone disease.

Cervical pachymeningitis may produce atrophic paralysis of the arms somewhat resembling that of progressive muscular atrophy: but in the former affection there is severe pain at the back of the neck and in the arms, the legs present symptoms of spastic paresis or paralysis, and sensory symptoms are present (*see* p. 384).

In idiopathic muscular atrophy and pseudo-hypertrophic paralysis the wasting does not begin in the small muscles of the hand; these muscles are usually spared; often more than one member of the family is affected; the disease commences before the patient reaches the age of twenty; fibrillary contractions are absent or at least occur exceedingly rarely; and the gait, and the manner in which the patient rises from the horizontal position on the floor, are often characteristic.

The diagnosis of progressive muscular atrophy from atrophy of muscles in "occupation paralysis" is considered on p. 251, and from the peroneal type of muscular atrophy on p. 258.

Pathological Anatomy.—A careful pathological examination has only been made in a few cases. The affected muscles are greatly diminished in size, their normal reddish colour is lost, they appear much paler than in health, or they are spotted with yellowish patches; in other cases their appearance is almost the same as that of fibrous tissue.

On microscopical examination, in some cases, even at an advanced stage, the condition of the muscles is one of simple atrophy—the transverse striation of the muscle fibres has been preserved, but the fibres are very narrow and the muscle nuclei of the sarcolemma are proliferated. In some cases the fibres present fatty and granular degeneration, and the transverse striation is lost. In other cases the changes are very marked, the muscle fibres have entirely degenerated, and the degeneration products having been absorbed, only the muscle sheaths remain. The muscle nuclei are greatly proliferated, the muscle sheaths contain only the proliferated muscle nuclei, and where these are absent the muscle sheath is collapsed (*see* Fig. 110).

Occasionally the muscle fibres present distinct longitudinal striation, and occasionally the muscle sheaths contain numerous round cells. The interstitial fibrous tissue may be increased in amount and often it contains fat cells.

The Spinal Cord presents no definite changes to the naked eye. On microscopical examination, the chief changes are in the anterior horns of grey matter. They are most marked in the cervical region, in the lumbar region they are often absent. Examination at a comparatively early stage has rarely been made; but in such cases (as in one examined by the writer in which death occurred from septic poisoning associated with an intercurrent disease) there is increased pigmentation of the nerve cells in the anterior grey horns of the cervical region; these nerve cells are shrivelled and granular, their processes are lost, and their nuclei may be indistinct.

At a later period the whole anterior horn has been found diminished

in size, the nerve cells of the anterior grey matter have completely disappeared, or only a few cells remain, presenting the degenerated appearances just described. The fine network of nerve fibres, normally seen in the anterior horns, has disappeared partially or almost entirely. The neuroglia connective tissue of this region is increased in amount, its nuclei are proliferated, and the small vessels are often dilated and their walls thickened.

At a comparatively early stage one or two groups of nerve cells of the anterior horns present degeneration, at an advanced stage all the cells have disappeared or have been markedly affected. Also it is stated that in comparatively early cases in an affected group of nerve cells,

FIG. 107.—Photograph of section of spinal cord (cervical region), progressive muscular atrophy. Weigert's stain. Note absence of degeneration in lateral pyramidal tracts. Anterior horns of grey matter small.

some cells present marked changes whilst adjacent cells are but little affected.

The white matter of the cord may be unaffected; but often there is atrophy of fibres in the antero-lateral columns just adjacent to the anterior horns. These are commissural fibres and the slight degeneration at this region has occasionally the appearance of a crescent-shaped zone around the anterior horns. The degenerated fibres are anterior to the crossed pyramidal tracts. In true cases of progressive muscular atrophy the crossed pyramidal tracts are quite normal in appearance (*see* Fig. 107).

In a few cases there has been slight degeneration in the column of Goll in the cervical region.

When the bulbar symptoms have been present the nerve cells of the motor nuclei of the medulla have presented degeneration similar to that seen in the nerve cells of the spinal cord (*see* description of amyotrophic lateral sclerosis).

The anterior nerve roots in the cervical region of the cord are atrophied. To the naked eye they appear diminished in size and thinner than the posterior roots. On microscopical examination marked degeneration of the nerve fibres has been seen, so that the nerve root has consisted of fibrous tissue and empty nerve sheaths, with here and there a nerve fibre. In the peripheral nerve trunks often only very slight changes have been detected, owing to the presence of so many normal nerve fibres from the posterior roots. Moreover the products of the complete degeneration of motor fibres have often become absorbed.

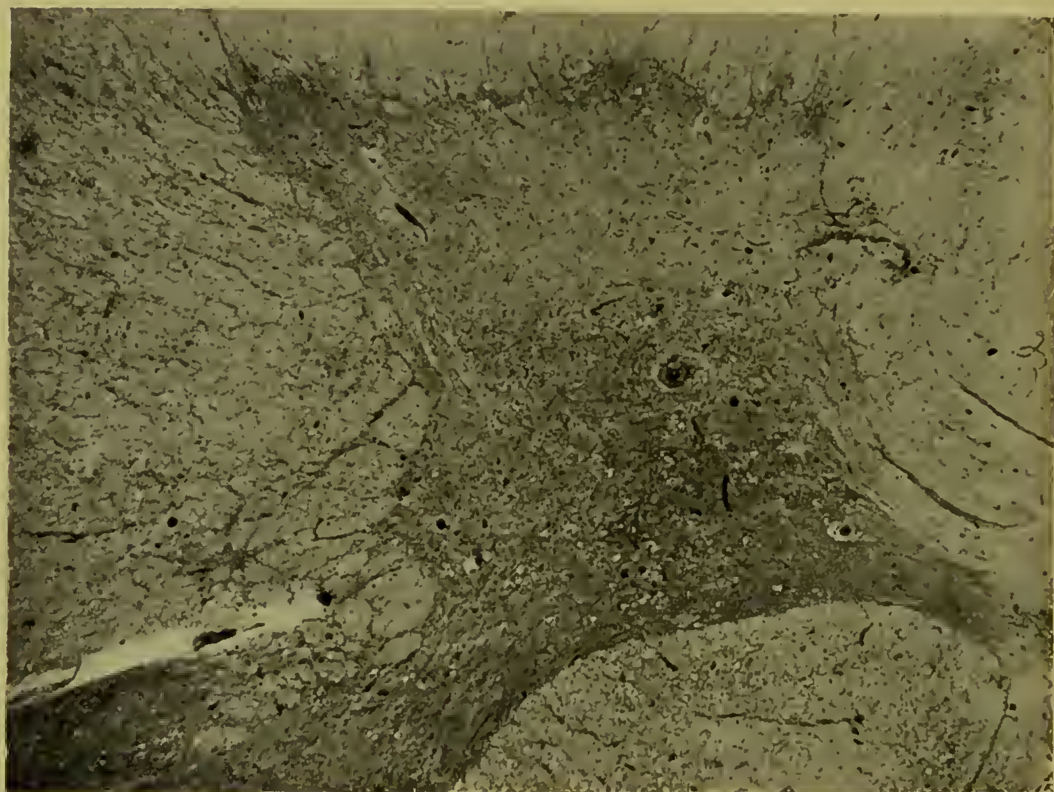

FIG. 108.—Microphotograph of Anterior Horn of Grey Matter, lower cervical region; progressive muscular atrophy. Nerve cells absent. Anterior horn at upper part, neck of posterior horn at lower part of photograph (aniline blue-black stain). Compare with normal anterior horn in Fig. 113.

The intra-muscular small branches of the motor nerves have sometimes presented marked degeneration : in other cases they have been reported to be normal.

The **treatment** is described along with that of amyotrophic lateral sclerosis.

As so many writers assert that all cases diagnosed as progressive muscular atrophy are really cases of amyotrophic lateral sclerosis or syringomyelia, I may briefly describe a case which appears to me to furnish clear evidence against this view. The patient was under the care of the late Dr. Leech, and on many occasions I had the opportunity of observing the distribution of the paralysis. Death occurred twenty-two years after the onset of the disease. Dr. Leech kindly gave me the

spinal cord for examination, and I have already published a report of the pathological changes.

I first saw the patient when a student in 1879. Both arms were then paralysed. I last examined the man, along with Dr. Leech, in 1898. Death occurred twelve months later.

The symptoms were those of slowly progressing muscular atrophy, with paralysis, affecting all the muscles of the hands and arms, and finally the muscles of the neck. For many years the arms hung helpless by the sides, and the head was bent forwards. There was no affection of sensation, and the bladder and rectum were not affected. There were no symptoms of bulbar paralysis. When I last examined him, more than twenty years after the onset of the disease, and only twelve months before his death, there was no spastic condition of the legs, the knee-jerks were present, there was no ankle-clonus, the patient could walk quite well, and the legs were unaffected. All the muscles of the arms were atrophied and paralysed. There was thus *no evidence*

I II

Fig. 109.—Anterior horns of grey matter (lower cervical), Weigert's stain. I. Normal, showing numerous fine nerve fibres and two nerve cells. II. Anterior horn in progressive muscular atrophy (same method of staining), nerve fibres very scanty, nerve cells completely degenerated.

of amyotrophic lateral sclerosis at the end of twenty years. The symptoms corresponded to those of progressive muscular atrophy of the Aran-Duchenne type, as described in the older text-books, and on pathological examination of the spinal cord I found that the condition confirmed this diagnosis. Pathologically, there was no evidence of amyotrophic lateral sclerosis. Briefly stated, the changes consisted in complete atrophy of the nerve cells of the anterior horns of grey matter in the cervical region (*see* Fig. 108). The fine nerve fibres in the anterior horns of grey matter in the cervical region had mostly disappeared, but in the posterior horns they were normal (*see* Fig. 109). The anterior horns of grey matter were smaller than in the normal condition. The

crossed and direct pyramidal tracts of the antero-lateral white matter presented *no sign of sclerosis or degeneration* (*see* Fig. 107).

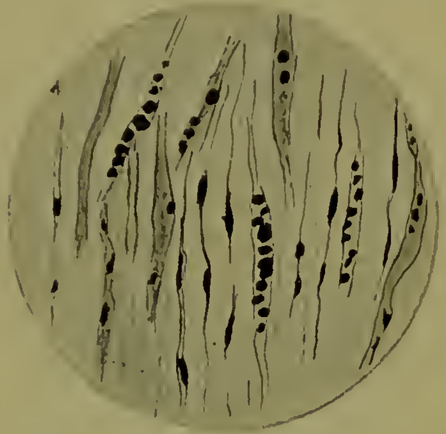

FIG. 110.—Muscle Fibres in Progressive Muscular Atrophy. Transverse striation lost. Muscle sheaths almost empty, containing simply the proliferated muscle nuclei.

The anterior nerve roots were markedly atrophied, the posterior were normal. The muscle fibres of the affected parts had entirely disappeared.

This case clearly demonstrates that there is a form of spinal muscular atrophy which may run its entire course without spastic paralysis or paresis of the legs developing, i.e. that there is a form of disease corresponding to the older description of progressive muscular atrophy (Aran-Duchenne type) which does not finally terminate in amyotrophic lateral sclerosis; that pathological changes in such cases may be localised to the anterior horns of grey matter; that the crossed pyramidal tracts may be quite normal; and that pathologically the lesion is a chronic anterior poliomyelitis. Both clinically and pathologically, therefore, the case differed from amyotrophic lateral sclerosis, and corresponded to the spinal progressive muscular atrophy of the Aran-Duchenne form, as described in the older text-books.

REFERENCES.

Williamson, R. T. *Lancet*, July 6, 1901 (reference to other cases given in this article) and *Edinburgh Medical Journal*, April 1907.

AMYOTROPHIC LATERAL SCLEROSIS

(Gr. ἀ. negative; μῦς, a muscle; τροφή, nutrition; σκληρός, hard.)

This affection was first described and separated as a definite disease by Charcot. It is a rare disease, which occurs usually in middle adult life; its exact cause is unknown, though the pathological anatomy has been carefully worked out.

Symptoms.—Briefly stated the symptoms, in a typical case, are: (1) Those of atrophic paralysis of the arms—resembling more or less the condition in progressive muscular atrophy; with (2) spastic paresis or paralysis of the legs; and (3) symptoms of chronic bulbar paralysis are often added.

The development of the disease is gradual. In most cases (but not in all), the first symptoms are those of weakness and atrophy of the arm muscles: at a later date, it may be 6 or 12 months afterwards, stiffness and weakness develop in the legs.

In the arms the affection usually begins in one hand : a little later the other is affected. At first there is weakness and wasting of the small muscles of the hands—thenar, hypothenar and interossei, as in progressive muscular atrophy (*see* Fig. 102). In many cases the loss of power is the first symptom and atrophy follows. It has been often pointed out, that at first there may be considerable loss of power quite apart from any atrophy, and that the weakness affects the whole extremity more diffusely than in progressive muscular atrophy. In other cases, however, the atrophy and weakness develop at the same time, and progress at the same rate, as in progressive muscular trophy. In the more rapidly progressive cases the loss of power and atrophy spread somewhat rapidly up the arms ; in the more chronic cases the weakness and wasting are localised to a few muscles at first and extend slowly from muscle to muscle. Atrophy is seen in the region of the thenar and hypothenar muscles and of the interossei, especially the first. In many cases the claw-like hand is produced at first, and then later the " monkey's hand " and the " skeleton hand." For description of these conditions, *see* p. 229. Also the same symptoms of weakness of the interossei can be detected as in progressive muscular atrophy (*see* p. 229, and Figs. 102 to 106).

The weakness and wasting spread up the arm, and in course of time the arms may become contracted or rigidly fixed in a definite position. When this occurs each arm hangs by the side of the body and any attempt to separate it is opposed by the action of the shoulder muscles. The forearm is semi-flexed and pronated ; it cannot be supinated without much force being employed and pain produced thereby. The wrists are semi-flexed and the fingers strongly flexed into the palm of the hand. (In many cases this marked contraction of the arm does not occur). As the atrophy increases the rigidity of the arms diminishes. The paralysis and atrophy spread to the neck muscles, the head is bent forwards, and the chin tends to rest on the sternum.

The legs gradually become rigid and weak ; and the symptoms of spastic paraplegia develop. There is rigidity of the legs to passive movement. The gait is spastic—the legs are kept stiff and extended at the knee in walking, the toes scrape the ground, and the advancing leg is thrown forward with a semi-circular movement at the hip at each step. At a later stage of the disease the leg muscles may become wasted, but they never atrophy so much as the arm muscles.

The knee-jerks are increased, often markedly, and patellar (rectus) clonus is often obtained. The tendo Achillis reflex is increased, and ankle-clonus is present. The plantar reflex is of the extensor type (Babinski's reflex). Oppenheim's reflex may also be obtained. In the arms the deep reflexes are increased ; the wrist-jerk (obtained by tapping the lower end of the radius), and the triceps-jerk are exaggerated. (Whilst in progressive muscular atrophy, as Strümpell points out, these deep arm reflexes are usually diminished or absent,

never increased. Sometimes clonic movements are produced by suddenly dorsiflexing the hand—hand-clonus.)

The jaw reflex is often increased, and sometimes a jaw-clonus may be obtained. This was first described by Beevor, who has never met with the symptom in any other form of disease, except once in hemiplegia. Beevor states that the best method of obtaining it is by placing "the finger transversely on the lower jaw just above the chin—or placing it on the teeth of the lower jaw, when the mouth is opened—and then tapping downwards on the finger, with a percussion hammer, or with the finger of the other hand. Another method is to place a paper knife on the molar teeth of one side and to tap it. If the reflex is increased a smart contraction of the masseters is produced, and in extreme cases a rhythmical clonus can be produced by keeping up the pressure down on the lower jaw." Beevor states that this symptom shows, that the sclerosis affects the pyramidal tract as high up as the level of the fibres going to the motor nucleus of the 5th cranial nerve.

(In the rare cases in which the leg muscles become atrophied the knee-jerks disappear.)

Fibrillary twitchings are present in the paralysed and atrophied muscles : they may precede any evidences of paralysis or wasting ; and may be the first sign of the disease.

On electrical examination there is a diminished excitability in the weak and atrophied muscles both to faradism and galvanism ; in other cases the partial reaction of degeneration is obtained ; and in very rare cases there is a complete reaction of degeneration in a few of the muscles.

There is no reaction of degeneration in the rigid leg muscles.

There is no form of anæsthesia at any period of the disease. Usually pain and other forms of sensory disturbances are absent : but occasionally there is numbness in the arms, through their fixed position at the side, and in very rare cases there is tearing pain in the limbs and neck.

The bladder and rectum are not affected.

At a late period of the disease symptoms of bulbar paralysis (glosso-labio-laryngeal paralysis) often develop. In some cases these symptoms appear at the onset of the disease, before the arms or legs are affected. The speech becomes slow, monotonous, and nasal ; the labial letters are badly pronounced ; the lips and tongue muscles become atrophied, and their movements gradually more and more impaired. Fibrillary twitching is seen in the affected muscles of the tongue and face. The movements of the soft palate and the pharyngeal muscles gradually fail. The soft palate cannot be raised on phonation ; there is increasing difficulty in swallowing, and fluids tend to regurgitate through the nose. The muscles supplied by the motor branch of the 5th nerve gradually become paralysed, and there is impairment in the movements of mastication and in the lateral movements of the jaw. The laryngeal muscles also become weak and are finally paralysed.

The lower facial muscles become more and more atrophied, the mouth becomes wide and its angles drop, and the naso-labial folds become more distinct. The face presents a melancholy expression ; the mouth remains half open, and saliva dribbles away from its angles. The patient is unable to pout the lips, to whistle, or to blow out a lighted match. Finally aphonia occurs from paralysis of the laryngeal muscles.

The ocular and upper facial muscles are unaffected, also the special senses are not impaired.

The patient becomes very emotional ; he laughs and cries without sufficient cause ; and though no marked intellectual defect is observed, he often becomes feeble-minded and childish. There is no loss of consciousness.

Finally the muscles of the trunk become affected and the patient is reduced to the most helpless condition, almost all the voluntary muscles of the body being affected more or less, except the ocular and upper facial muscles. Attacks of tachycardia may occur. The breathing may become very difficult from affection of the respiratory muscles. Food is apt to pass into the larynx owing to the bulbar paralysis.

Death may occur from broncho-pneumonia, excited by food particles passing into the air passages, or from asphyxia owing to paralysis of respiratory muscles, or from inanition. In other cases intercurrent diseases such as pneumonia, phthisis, or bronchitis are the cause of the fatal termination.

The duration is usually from 1 to 3 years. (In 38 out of 42 cases the duration was under 4 years—Probst.)

Course.—In most cases the arms are first affected, then the legs and finally bulbar symptoms may appear. In some cases the legs are first affected with spastic paralysis, and the other symptoms develop later. In a third rare group of cases bulbar symptoms develop first and then the arms and legs are affected.

Often one arm or one leg is affected more than the other at first ; and sometimes one arm or one side is affected for several months before the other. In 6 or 8 months frequently there is marked affection of both the arms and legs.

There are three modifications of the disease which require brief mention :—

1. Rare cases are on record, in which spastic symptoms fail, and clinically the condition is one of atrophic paralysis, with final bulbar symptoms ; and yet post-mortem examination reveals, in addition to the change in the anterior horns of grey matter, degeneration of the lateral columns and the other changes of amyotrophic lateral sclerosis. The cases present clinically the symptoms of progressive muscular atrophy with bulbar paralysis : whilst pathologically the changes are those of amyotrophic lateral sclerosis.

2. In other rare cases, the atrophic paralysis is very slight, but the spastic paralysis marked ; pathologically there are only slight

changes in the anterior horns, but marked changes in the lateral columns.

3. Senator has recorded a case in which the symptoms were those of amyotrophic lateral sclerosis, but pathologically there were no changes in the lateral columns.

Pathology.—The pathological anatomy of the disease has been worked out very thoroughly. It has been shown, that amyotrophic lateral sclerosis is a disease affecting the cortico-motor conducting tract on both sides, and sometimes the whole of this tract; or in other words it is a disease affecting both the upper and lower (central and peripheral) motor neurons. In some cases, as in that recorded by Mott (*Brain*,

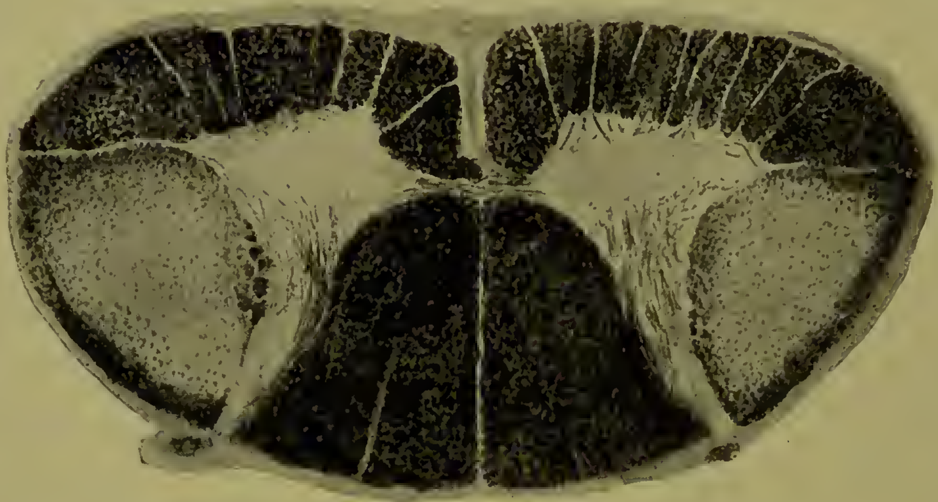

FIG. 111.—Section of the Spinal Cord in the lower cervical region in a case of amyotrophic lateral sclerosis, Weigert's stain: normal nerve fibres black, degenerated parts pale. Note pale degenerated area in each lateral column. Degeneration affects the crossed pyramidal tracts, and *also* extends to antero-lateral white matter just in front of this tract (*see* text). In the anterior horns of grey matter the fine nerve fibres are absent, whilst fine normal fibres are seen running from the posterior horn to the median grey matter. (Under the high power of the microscope the nerve cells of the anterior horns were found to be absent or atrophied.)

1895, p. 21), there was marked degeneration of the motor path from the brain cortex to the periphery: there was degeneration of the large pyramidal cells and nerve fibres of the motor cortex, of the motor fibres of the internal capsule, crus, pons, medulla, and crossed pyramidal tracts of the cord, of the nerve cells of the anterior horns of grey matter, and of the fibres of the ulnar nerve.

Schmaus and Sacki point out, that, in addition to the lesion of the upper and lower motor neurons, there is also an affection of the neurons in the cord which connect the motor neurons of various levels with each other (neurons which furnish commissural fibres in the antero-lateral columns).

In the spinal cord the trophic centres of the atrophied muscles, and the motor tracts are degenerated, i.e. nerve cells of the anterior

horns, especially in the cervical region, and the lateral pyramidal tracts.

The *grey matter* of the anterior horns is markedly affected in the cervical region. The ganglion cells are partially degenerated, or they have altogether disappeared (simple or pigmental atrophy of nerve cells). Nissl's stain shows that the chromatophile bodies lose their angular form and present a fine dustlike appearance. Pigment is collected around the nucleus which is often displaced laterally. The cells may be transformed into a small deeply stained mass. The fine network of nerve fibres of the anterior horns has partially or completely disappeared. The neuroglial interstitial tissue is proliferated, its

FIG. 112.—Microphotograph of Anterior Horn of Grey Matter, lower cervical region, amyotrophic lateral sclerosis. Nerve cells absent. Compare with Fig. 113.

nuclei are increased in number, and it contains star-like Deiter's cells. The changes in the network of fine nerve fibres extend as far as the base of the posterior horn. The grey matter is more degenerated at the middle of the anterior horn than at its periphery. The reflex collateral fibres are usually spared, and, as shown in the accompanying Fig. 111, normal fine nerve fibres are seen passing from the posterior horn forward towards the centre of the grey matter; whilst the fine network of nerve fibres in the anterior grey matter has completely or partially disappeared. Sometimes the anterior horns of grey matter appear smaller than normal. In the lumbar region the changes are usually very slight or much less marked than in the cervical region.

The walls of the blood vessels may be thickened.

In the *white matter* of the cervical region of the cord there is marked degeneration in the lateral or crossed pyramidal tracts. Frequently there is a diffuse slight degeneration in the surrounding parts of the antero-lateral columns—between the anterior horns and the surface of the cord—but this degeneration is less marked than in the crossed pyramidal tracts, and it does not usually affect the direct cerebellar tract and the ascending antero-lateral tract of Gowers. The portion of the anterior lateral region known as the intermediate lateral bundle is slightly degenerated. Sometimes the lateral limiting layer (close to the lateral grey matter) is affected; sometimes it appears almost

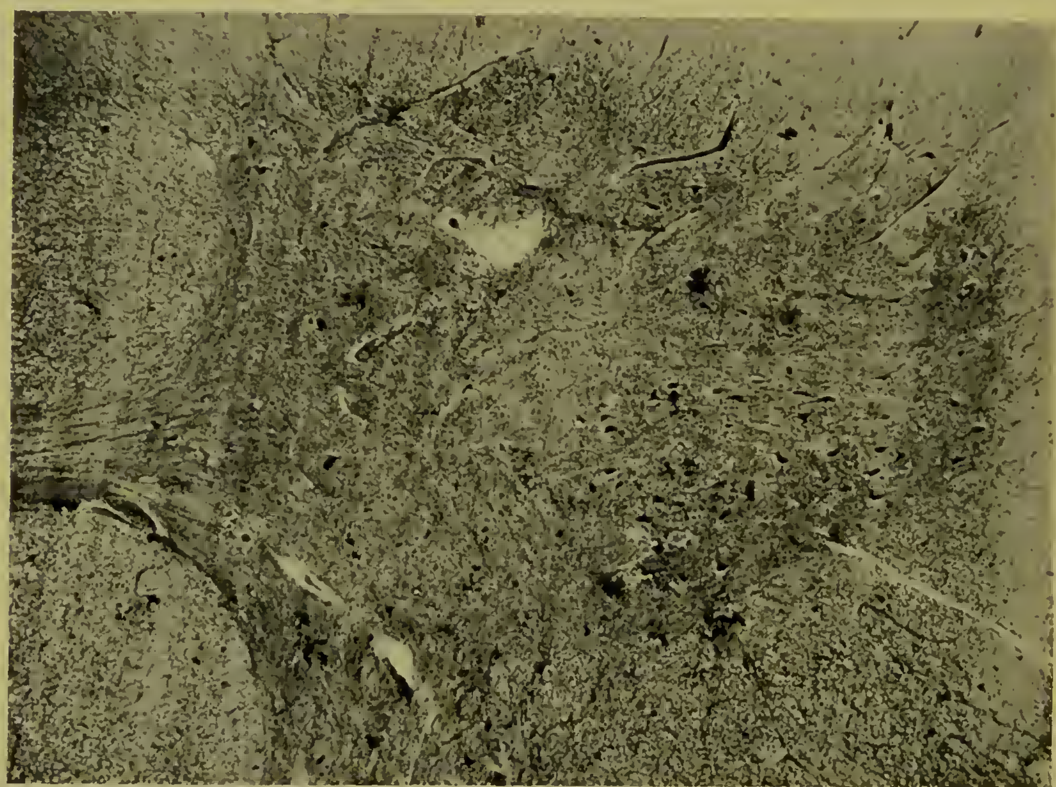

FIG. 113.—Microphotograph of Normal Anterior Horn of Grey Matter, lower cervical region. Nerve cells numerous and distinct (black). Stained in same manner as Fig. 112 (carmine).

normal. In some cases the anterior commissure is degenerated. Frequently the direct pyramidal tracts are degenerated in the cervical region. In the lumbar region, the degeneration in the white matter is limited to the crossed pyramidal tracts. The posterior columns are usually normal, but slight degeneration has been found in the columns of Goll, in several cases recorded. The columns of Clarke are usually not affected.

Degeneration is found in the anterior nerve roots in the cervical region, and in the roots of the motor nerve arising from the medulla. In the peripheral motor nerve trunks changes are less distinct, and sometimes no change has been detected. The small intra-muscular nerve fibres have been found markedly degenerated.

The atrophied muscles are pale red, reddish yellow, or yellow in colour. The muscles of the hand, arm, and tongue are most atrophied. The muscle fibres are narrow, and their nuclei increased; they may present fatty degeneration, or loss of transverse striation. Amongst the atrophied fibres sometimes very large fibres are seen.

In the medulla there is degeneration of the anterior pyramidal tracts, but this is less marked than in degeneration from a cerebral lesion. The degeneration of the motor fibre may cease at the medulla; in other cases it can be traced to the pons and crus; in some cases to the internal capsule and cerebrum. In sections stained according to Marchi's method, degeneration of the posterior longitudinal bundle of fibres has been found in the medulla.

The motor nuclei of the medulla—hypoglossal, facial, vagus accesorius and motor nucleus of the 5th nerve—present atrophy, disappearance of ganglion cells, and other changes similar to those in the grey matter of the anterior horn.

The cortex of the motor area of the brain, in some cases, presents no change. In other cases changes are found in the nerve fibres and nerve cells of this region (Kojewnikoff, Charcot, Marie, Mott, Spiller). These changes have been discussed by v. Czyhlarz and Marburg, who conclude that it has only been definitely proved that the radial fibres of the motor cortex and the nerve cells in relation therewith are degenerated. They believe that the fibres degenerate first, and the cells afterwards. Atrophy of the large pyramidal cells of the motor area of the cortex has been recorded, and their number has been found smaller than in health. Mott points out that in the cortex the degeneration is confined to the large Betz cells; the smaller and medium-sized pyramids, and the layers of tangential and super-radial fibres, being quite unaffected. Probst and Spiller have found degeneration in the corpus callosum. Spiller has employed the degeneration of the cortex to map out the motor region of the brain.

The degeneration of the white matter in the cord presents the usual appearance of sclerosis, similar to that seen in secondary degeneration. The degeneration of nerve fibres is primary, and the proliferation of connective tissue is secondary. The degeneration of the motor fibres of the lateral pyramidal tracts appears to begin below and to ascend (Schukowski).

Thickening and sclerosis of the vessels in the degenerated parts is sometimes found, but this is a secondary change. The changes in the cord and brain in amyotrophic lateral sclerosis are bilateral, though they are not always of equal intensity on the two sides.

Probst has made an analysis of 53 cases of amyotrophic lateral sclerosis, including 11 of his own cases. Of these 53 patients, 26 were males and 27 females; 10 patients were between 23 and 30 years of age; 16 between 30 and 40; 11 between 40 and 50; 13 between 50 and 60; 5 between 60 and 70; and 1 between 70 and 80. As regards the duration of the

disease, in 9 cases it was less than 1 year; in 16 cases it lasted 1–2 years; in 9 cases 2–3 years; in 4 cases 3–4 years; in 4 cases longer than 4 years. In 55 cases the disease began 20 times in an upper extremity; 9 times in the lower limbs; 5 times in all extremities about the same period; 9 times the first symptoms were bulbar; 9 times a hemiplegic form was seen at the onset. Usually bulbar symptoms were seen early; in a few cases they did not appear for a year; in one case not for two years. In a few cases bulbar symptoms did not develop.

The cause of the degeneration of the motor neurons in amyotrophic lateral sclerosis is not known. Some writers regard it as the result

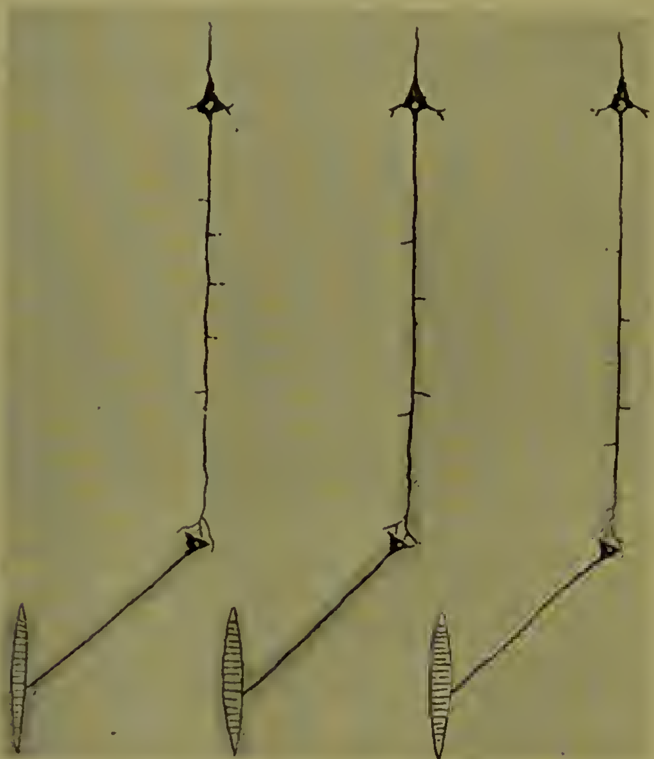

Fig. 114.—Upper and lower motor neurons and muscles. Degenerated portions of neurons and degenerated muscles are shaded. Neurons in progressive muscular atrophy to the left; in amyotrophic lateral sclerosis in the centre; in primary lateral sclerosis to the right.

of an abnormal congenital tendency or defect in these neurons. Others think it is due to a chronic intoxication. Excessive functional activity of the motor neurons (overwork), associated with insufficient feeding or weakening disease, is suggested as a possible cause by v. Czyhlarz and Marburg. Collins has shown that the disease occurs most frequently amongst the "working classes," and that a number of cases have developed shortly after parturition.

Diagnosis.—Though *progressive muscular atrophy and amyotrophic lateral sclerosis* are probably closely allied, and possibly forms of one pathological process, still the results of the microscopical examination in a few cases, such as that which I have described on p. 237, furnish

clear evidence that there are two separate diseases, or two distinct forms of one affection, to which the names of amyotrophic lateral sclerosis and progressive muscular atrophy may be respectively applied.

The diagrams (Fig. 114) demonstrate the localisation of the changes. The upper and lower motor neurons and muscles are indicated. The upper motor neuron is the nerve cell in the motor cortex of the brain

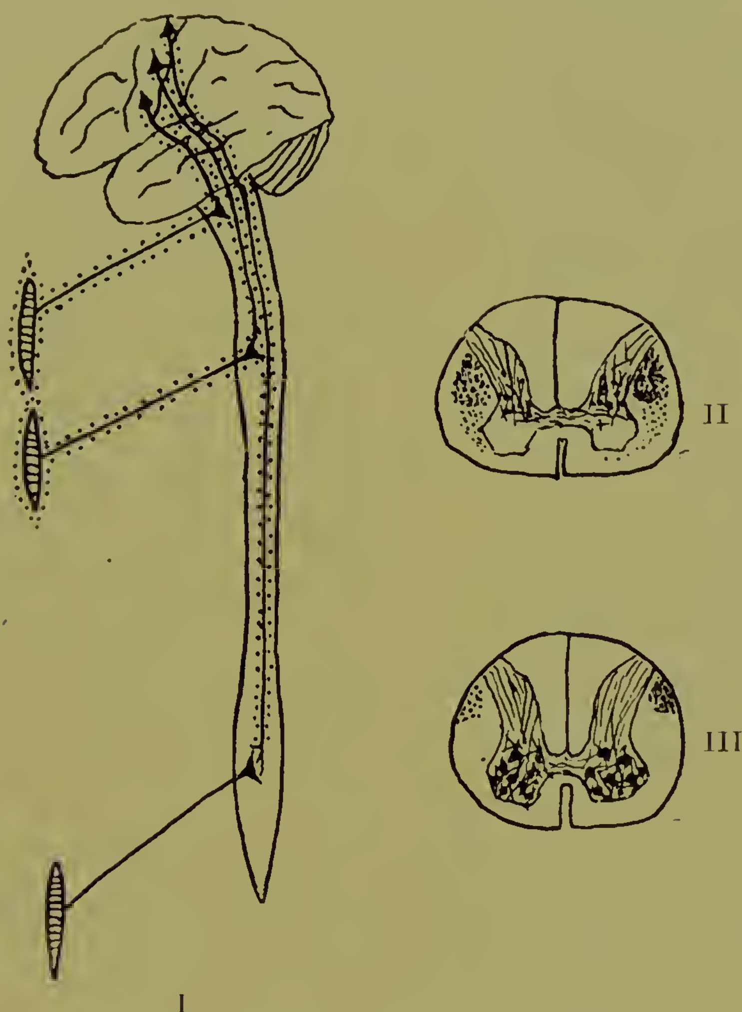

FIG. 115.—Diagrams showing seat of lesions in amyotrophic lateral sclerosis.

I. Brain and cord showing upper and lower motor neurons for arm, leg and bulbar muscles. Neurons which are diseased are marked with dots.

II. Cervical region of cord: degeneration (marked by dots) of crossed pyramidal tracts and slight degeneration anterior to these tracts. Absence of nerve cells and fine nerve fibres in anterior horn of grey matter.

III. Lumbar region of cord: degeneration in crossed pyramidal tracts (marked by dots). Cells and fine nerve fibres in anterior horns normal.

and its axon passing down in the crossed pyramidal tracts of the spinal cord. The lower motor neuron is the nerve cell in the anterior horn of grey matter and its axon passing into the motor nerve root and thence to the muscles. Now, in progressive muscular atrophy the degeneration in the nervous system is localised to the lower motor neurons—the cells in the anterior horns and the nerve fibres proceeding therefrom.

In amyotrophic lateral sclerosis the lower motor neuron is degenerated in the same manner in the cervical region; but in addition the upper motor neuron is affected, the degeneration involving the axons of these neurons in the cord. Sometimes the degeneration extends up to the medulla, the internal capsule, or even to the motor cells in the cortex. The lower motor neuron is not affected in the lumbar region (*see* Fig. 115), or affected only at a late stage of the disease.

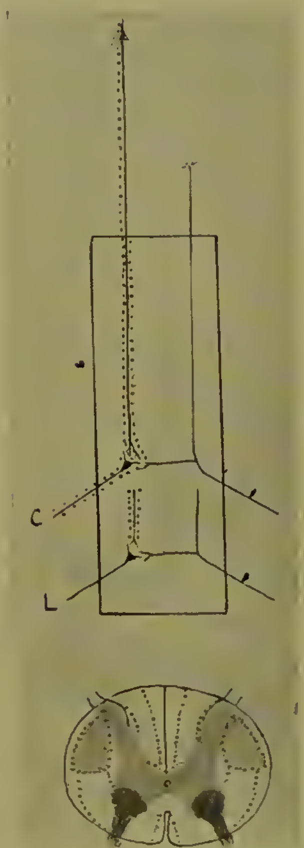

FIG. 116.—Lower figure = transverse section of Spinal Cord, cervical region. Amyotrophic lateral sclerosis. Shaded parts (lateral pyramidal tracts and anterior horns) are degenerated. Upper figure = antero-posterior section of Spinal Cord. To the left is indicated the upper motor neuron (cell in the motor cortex of brain with long fibre in lateral pyramidal tract), and the lower motor neuron (cell in anterior horn, fibre in peripheral nerve). Lower motor neuron in cervical region = C, in lumbar region L. To the right is indicated the sensory neuron at the posterior part of the Cord—posterior root, ascending fibre, and collateral, the latter running transversely to the nerve cell of anterior horn. The neurons marked with dots are degenerated in amyotrophic lateral sclerosis.

Often the clinical diagnosis of progressive muscular atrophy can only be regarded as a probable one, since cases which at first have presented the features of this disease have developed the symptoms of amyotrophic sclerosis at a later period; and in rare instances an affection which has appeared to be due to progressive muscular atrophy during life has proved to be amyotrophic lateral sclerosis on pathological examination.

The paralytic condition of the arms is similar in many respects in both diseases; but the following are important points in the differential diagnosis of the two affections :—

1. In favour of amyotrophic lateral sclerosis would be a spastic condition of the legs—spastic gait, scraping of the toes in walking, rigidity of the legs on passive movements, increased knee-jerks, ankle-clonus, and the extensor type of plantar reflex, Babinski's reflex. These symptoms are all absent in true progressive muscular atrophy.

2. Symptoms of bulbar paralysis would be in favour of amyotrophic lateral sclerosis. According to Dejerine, bulbar symptoms do not occur in true progressive muscular atrophy.

3. Amyotrophic lateral sclerosis, as a rule, runs a more rapid course than progressive muscular atrophy. The duration of the disease in progressive muscular atrophy is often very chronic — ten, fifteen, or twenty years; whilst in amyotrophic

lateral sclerosis the disease usually terminates fatally in from one to four years, though a few exceptional cases of longer duration have been recorded.

4. The increase of the wrist-jerk is in favour of amyotrophic lateral sclerosis : loss of this reflex is in favour of progressive muscular atrophy.

5. Paresis or paralysis in muscles which are not yet definitely atrophied is in favour of amyotrophic lateral sclerosis and against progressive muscular atrophy.

The spastic condition of the legs and the increased reflexes separate amyotrophic lateral sclerosis from idiopathic muscular atrophy and from cases of muscular atrophy due to *peripheral neuritis.* In *idiopathic muscular atrophy* the age at the onset is under 20 ; the atrophy begins in the shoulder and upper arm muscles ; the small muscles of the hand are not affected ; fibrillary contractions are almost always absent ; there is a tendency for more than one member of a family to suffer ; the facial expression and gait are quite different (" myopathic face," waddling gait, etc.).

In cases which clinically correspond to the description of " *primary lateral sclerosis* " there is no atrophy of muscles of the arms or other parts ; bulbar symptoms are absent.

Syringomyelia in the cervical region may cause atrophic paralysis of the arm and spastic paraplegia ; but sensation is affected also in most cases (analgesia and thermo-anæsthesia being present). If the posterior horns of grey matter are not affected in syringomyelia sensory symptoms may be absent ; but the course of the disease is much longer than in amyotrophic lateral sclerosis.

Cervical myelitis, cervical caries, and cervical pachymeningitis may cause atrophic paralysis of the arms and spastic paraplegia ; but in addition, diminution of sensation (or anæsthesia) and bladder symptoms are usually present ; also there are no bulbar symptoms and no fibrillary contractions. Moreover, in cervical caries there are the bone symptoms of vertebral disease, and in cervical pachymeningitis there are severe pains in neck and arms before the onset of the paralysis.

In very rare instances multiple sclerosis has caused spastic paralysis of the legs with atrophic paralysis of the arms ; but usually nystagmus, scanning speech, and intention tremors, have been present.

Treatment of Progressive Muscular Atrophy and Amyotrophic Lateral Sclerosis.

The only treatment which has been of distinct service in these diseases is the hypodermic injection of strychnine.

Sir Wm. Gowers and Dr. J. Taylor state that " the evidence of arrest consequent upon this treatment is now undoubted, and it should be remembered that the hypodermic administration of the drug has been successful in cases in which it has failed to affect the diseases when

given by mouth." They recommend one injection daily, and prefer the nitrate of strychnine to other salts of the alkaloid. The dose at first should be $\frac{1}{100}$ of a grain quickly increased to $\frac{1}{40}$ or more. When the disease is apparently arrested the drug should be omitted for one week in three or four. The injection can be made at any convenient place.

Writing in 1891 Sir Wm. Gowers states that in seven almost consecutive cases in middle life this treatment has been followed by arrest within a month of its commencement, and the arrest has been permanent in all the cases but one. In senile cases the treatment failed.

Collins states that "to a certain extent" his experience corroborates the opinion of Gowers. He has had under his observation for eight years a patient in whom the atrophy seems to have remained at a standstill after such treatment combined with the use of faradic electricity, massage, and general hygienic measures. He has treated two other patients in the same way "with encouraging results." But in several other cases the drug has failed. Collins recommends the dose of the nitrate of strychnine to be gradually increased from $\frac{1}{80}$ to $\frac{1}{16}$ or $\frac{1}{8}$ of a grain (hypodermically) according to the results, and the injections to be continued from two to four months. Other physicians have reported that they have not obtained definite results from the treatment. As no other drug has been shown to have any influence on the disease, it is only fair to the patient that this method of treatment should be tried in all cases at first.

In two cases of amyotrophic lateral sclerosis with marked progressive atrophy of the arm muscles, I have recently given a fair trial of the hypodermic injections of strychnine without definite results. The disease advanced and terminated fatally.

The occupation should be discontinued. Massage is of service according to Collins, but he recommends that only the "gentlest kneading movements should be employed." Passive movement should be made with the object of preventing deformity. Collins thinks that electricity is of some service in delaying the progress of the disease. A very weak faradic current should be applied daily to the affected muscles for about five minutes; a strong current should be avoided. When there is no response to faradism, the galvanic current should be employed.

Thyroid and other animal extracts have been shown to be useless in nearly all cases. Oppenheim, however, states that in one case thyroid preparations were of service (no details given). Everything should be done to improve the general health and to prevent complications.

In amyotrophic lateral sclerosis, when bulbar paralysis is present, there is great difficulty of swallowing, and it may be necessary to feed the patient with the œsophageal tube.

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Muscular Atrophy from Over-use; Professional or Occupation Paresis

Cases of muscular atrophy are occasionally met with owing to previous over-use of the wasted muscles. The small muscles of the hand, especially the thumb muscles, are most frequently affected. This form of atrophy has been observed in file-cutters, laundry women, who iron linen for many hours daily, basket-makers, locksmiths, joiners, etc.

The atrophy is usually unilateral, and there is sometimes paræsthesia or diminution of sensation in the sensory distribution of the affected nerve areas, but in other cases sensory symptoms are entirely absent.

The wasting is probably due in some cases to pressure on the muscles, or on the nerves which supply them: in other cases it is probably due to changes in the motor nerve cells in the anterior grey matter.

The wasting is localised and shows no tendency to extend to other muscles, even after many years. When the over-use of the muscles is discontinued, there is no increase of the atrophy, and often it disappears.

The **diagnosis** can be made by the history of over-use of the muscles, and by the fact that even after many years the atrophy does not extend to other muscles. Also the reflexes are normal, and other signs of amyotrophic lateral sclerosis and progressive muscular atrophy are absent.

Paræsthesia or diminution of sensation, if present, in the atrophied region would be evidence against progressive muscular atrophy and amyotrophic lateral sclerosis. On changing the occupation there is no further increase of the atrophy in cases of occupation paralysis; whilst in progressive muscular atrophy and amyotrophic lateral sclerosis the wasting progresses in spite of the change of work.

In one of my own cases the atrophy was limited to the small muscles of the thumb, and affected only those supplied by the median nerves. At the end of five years it had remained localised to these muscles, and there were no signs of amyotrophic lateral sclerosis or progressive muscular atrophy.

Treatment.—The use of the affected muscles should be discontinued: rest is important: galvanism and tonics such as strychnine may be used.

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SYRINGOMYELIA AND SPINAL GLIOSIS

(Greek—*σῦριγξ*, a tube; *μυελός*, marrow.)

These two conditions are described together because they cause similar symptoms and are often associated pathologically. Both conditions may produce muscular atrophy.

In spinal gliosis there is a localised formation of new neuroglial tissue in the grey matter of the spinal cord.

Cavities may arise in the cord by degeneration of this new formed glial tissue, or through developmental defects in connexion with the central canal, or in other ways. The conditions in which cavities are present in the cord are known as syringomyelia.

Pathological Anatomy.—The external appearance of the cord in spinal gliosis or syringomyelia is often unaltered, but in other cases the cord is swollen in the region of the lesion. When a cavity is present, syringomyelia, the cord may feel like a tube to the finger. On section a large cavity may be seen and clear serous fluid may escape. In other

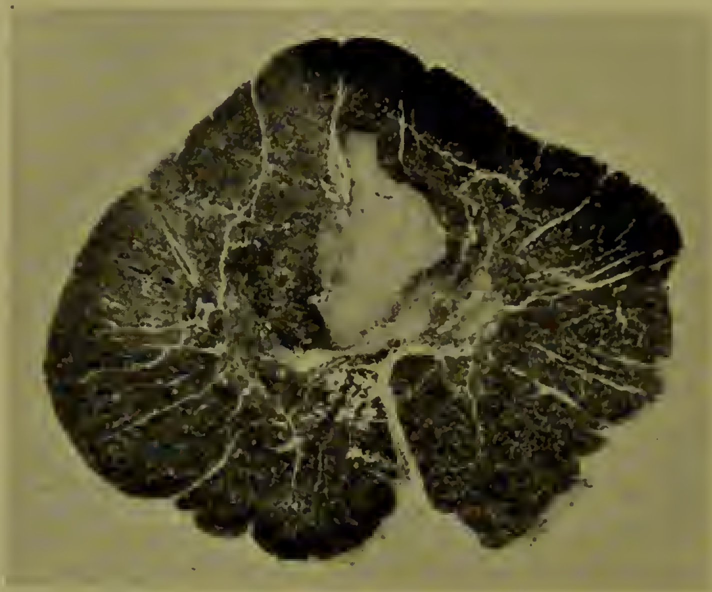

Fig. 117.—Section of Spinal Cord. Weigert's stain. Syringomyelia. Cavity in the posterior columns.

cases there is a small cavity in the region of the central canal or posterior to this region. In many cases, gliosis, no cavity can be seen, but there is a tumour mass in the centre of the cord or in the posterior grey matter which extends longitudinally for a considerable distance. The tumour may occupy the cervical region or both cervical and dorsal regions; occasionally it extends to the medulla. Gliosis or syringomyelia is chiefly met with in the cervical and upper dorsal regions of the cord. The new formed tissue in gliosis consists of neuroglia cells and fibres. The cavities in syringomyelia are surrounded by a thick zone of new formed glial tissue, and are often lined with a distinct firm sinuous membrane (*see* Fig. 119); sometimes a distinct epithelial lining of ependyma cells can be detected on the interior of this membrane at some parts. This neuroglial tissue contains no nerve fibres, and is simply stained brown in specimens prepared according to Weigert's hæmatoxylin method. Weigert has pointed out that the neuroglia tissue around the cavities consists almost entirely of

glial fibres with few cells, and that it does not resemble the structure of a glioma. Secondary descending degeneration may occur in the pyramidal tracts, and ascending degeneration in the direct cerebellar tracts and in the columns of Gowers. Also in the posterior columns gliomatous proliferation may be present in the ventral regions; close to the posterior commissure as a wedge-shaped area along the median septum, and in a small bundle between the columns of Goll and Burdach. The latter begins at the central gliomatous proliferation, and extends along the anterior two-thirds of the posterior columns.

The cavities are chiefly found in the posterior part of the cord, and not infrequently are situated in the posterior horn of grey matter

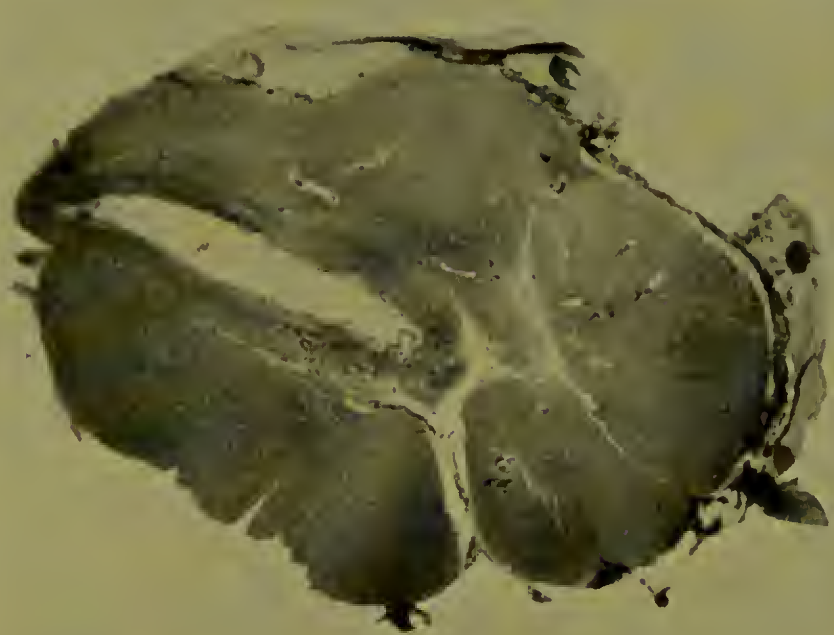

Fig. 118.—Spinal Cord. Syringomyelia. Note cavity in posterior grey matter to left of figure.

(*see* Fig. 118). Sometimes almost the whole of the grey matter is destroyed by a cavity.

Causation of syringomyelia.

(*a*) In the simplest forms the cavity in the cord is a dilated central canal; the condition is known as hydromyelia, and it is probably a *congenital* affection. Such congenital cavities are lined by ependyma cells, and surrounded by a layer of neuroglia.

(*b*) In other cases the cavity is due to a defect in the development of the cord—a defect in the closure of the central canal.

In the development of the spinal cord there is first a dipping down of the epiblast of the embryo. This forms the primitive furrow of the embryo. The anterior wall of this furrow becomes thickened to form the white commissure and anterior part of the grey commissure. The lateral and anterior columns develop, and then the posterior columns.

As the posterior columns increase in thickness the posterior part of the canal becomes narrowed, and its walls unite near the anterior end to form the posterior grey commissure, and so the tube is divided into two parts. Later the posterior part of the canal is closed.

When this closure does not occur the simplest form of syringomyelia is produced. Such cavities have been found in new-born children.

In adults cavities may be regarded as congenital, when they are lined with ependyma cells and situated in a median position or in the position of the central canal, and when other spinal changes are absent.

FIG. 119.—Cavity in Spinal Cord. Syringomyelia. Note sinuous membrane bounding cavity and layer of neuroglia tissue external to it. (From the same section as Fig. 118, but more highly magnified.)

There are other causes for cavities which are not lined with ependyma cells.

(*c*) In some cases of syringomyelia there is a primary gliosis, and the cavities are formed by the degeneration and breaking down of the new formed gliomatous tissue. In very rare cases degeneration in a sarcomatous growth may produce a cavity.

(*d*) Cavities may be formed in the cord which resemble those of syringomyelia, owing to the degeneration of diseased tissue in spinal hæmorrhage, and in spinal softening from vascular changes and other causes. In spinal syphilis a softened area of the cord, caused by disease of the spinal vessels, sometimes breaks down and a cavity is formed. Also cavities resembling those of syringomyelia have been found associated

with pachymeningitis and chronic meningitis. (This group of cases should be separated from true syringomyelia.)

It has been frequently observed that syringomyelia has followed injury to the back, and it is conceivable that injury may be the starting-point of the development of new glial tissue. Minor believes that a central hæmatomyelia, caused by injury, is not infrequently the starting-point of syringomyelia. Possibly a spinal hæmorrhage occurring at birth may also be the starting-point of certain cases of syringomyelia.

Schlesinger thinks that median cavities lined by ependyma cells may be attributed to developmental abnormalities. Lateral cavities may be caused in other ways: in some cases they are due to gliomatous proliferation and degeneration, in other cases to vascular disease and œdematous or inflammatory processes.

Symptoms.—Syringomyelia has been occasionally found post mortem, when there have been no definite symptoms during life. But in many cases when the pathological changes are in the common situation, in the cervical enlargement of the cord, the following group of symptoms is met with :—

1. Loss of sensation to pain and temperature in the arms and trunk, whilst tactile sensation is preserved.
2. Progressive muscular atrophy in the arms.
3. Trophic and vaso-motor symptoms.
4. Frequently spastic paresis or paralysis of the legs.

The onset of the symptoms is very gradual. The sensory changes are said to appear first. The sensation for pain (pin prick, faradic current, etc.), and the sensation for heat and cold are lost or diminished; whilst the sensation for tactile impressions, for pressure and the muscular sense (for position) are preserved. This is described as dissociated anæsthesia, and is the common and typical sensory affection of syringomyelia. Loss of the vibrating sensation has been observed in a few cases.

(In some cases the sensation for heat alone is lost, whilst cold is distinguished; in other cases the reverse condition is found; whilst in others a moderate degree of warmth and cold are distinguished correctly, but very hot or very cold objects are not correctly distinguished.) The sensory symptoms are noted more frequently in the arms than in the legs.

The analgesia and thermo-anæsthesia do not follow the distribution of separate nerves, but in the arms and trunk correspond to the regions supplied by posterior spinal nerve roots. The sensory symptoms are usually present in the parts affected with muscular atrophy. Occasionally in the legs there is Brown-Séquard's paralysis. Paræsthesia is often present. Pains sometimes occur in the limbs and back.

The muscular atrophy begins in the hands, seldom in the upper arm or shoulder muscles. The small muscles of the hands undergo

gradual atrophy, the interosseous spaces become depressed, and the thenar and hypothenar eminences waste. A claw-shaped hand gradually develops, as in progressive muscular atrophy. The atrophied muscles show fibrillary contractions. On electrical examination the reaction of degeneration is occasionally met with in some muscles or in portions of an atrophied muscle. Sometimes there is simple diminution of electrical excitability, in other cases no definite electrical changes can be detected. The atrophy is usually better marked in one hand than the other. The atrophied muscles become weak and finally paralysed. The wasting extends, and, as the muscles affected at first are chiefly those supplied by the median and ulnar nerves, there is a tendency for over-action of the extensor muscles—a tendency for the hand to become extended at the wrist and finger joints and to assume the position known as the "preacher's hand."

The legs are very rarely atrophied. In such cases the disease has extended to the lumbar region of the cord.

The legs often become spastic (spastic paresis or paralysis), the knee-jerks become increased, and ankle-clonus is present. The sphincters are sometimes paralysed, in other cases unaffected.

Owing to weakness of the trunk muscles there is often lateral curvature of the spine.

Usually the symptoms of the disease are bilateral.; in exceptional cases unilateral.

Frequently there are trophic changes in the limbs. The skin may become glossy, or thick and horny. Skin affections such as pemphigus, herpes zoster and urticaria are met with. Frequently bullæ form on the skin (hands), which leave troublesome ulcers behind. The nails may become fissured and cracked, or they may fall off. The hands are often bluish or red, and sometimes œdematous. Whitlows, thickening of the terminal phalanges, ankylosis of the finger joints, and mutilation of the phalanges are sometimes observed.

The hand is sometimes enlarged—"main succulente" of Marinesco. The chief change is a diffuse swelling, especially on the back of the hand.

Unilateral sweating has been occasionally observed.

The bones may become thickened or brittle, and joint changes similar to those of tabes may occur; but it is stated that the changes are usually confined to the joints of the arms. Occasionally the cranial nerves are affected (owing probably to extension of the changes to the medulla and pons). Thus paralysis of one vocal cord, of the soft palate, of the tongue, and of the face have been noted. Analgesia and thermo-anæsthesia or other sensory symptoms in the distribution of the fifth nerve have been occasionally observed. Difficulty in swallowing and affections of the cardiac and respiratory functions have been recorded.

The pupils and palpebral fissures are often diminished on one side (the side on which other changes are most marked), or the pupils and palpebral fissures are small on both sides. The pupils react to light,

but may dilate imperfectly in the shade. Concentric restriction of the field of vision and hysterical symptoms are sometimes present. Nystagmus is occasionally observed.

Ataxia is occasionally noted, and the symptoms may then somewhat resemble those of tabes.

Atypical cases occur in which the usual sensory symptoms are present but muscular atrophy absent; in other cases there is tactile anæsthesia as well as analgesia and thermo-anæsthesia; cases are also on record in which muscular atrophy is the only marked symptom.

A number of cases of unilateral syringomyelia have been published. In some of these only one posterior horn of grey matter has been affected, and clinically analgesia and thermo-anæsthesia have been present on the side of the lesion.

The course of the disease is very chronic. Death occurs from exhaustion, from impairment of the function of the medullary centres, from complications such as bed-sores, cystitis etc., or from intercurrent disease.

The **diagnosis** of syringomyelia is easy in typical cases; very difficult or impossible in atypical cases. If the sensory disturbances of syringomyelia are absent, a certain diagnosis cannot be made. Amyotrophic lateral sclerosis, progressive muscular atrophy and anterior poliomyelitis may all cause atrophy of hand and arm muscles similar to that of syringomyelia; but in these conditions there is not the loss of sensation to pain and temperature which is characteristic of syringomyelia. The atrophic paralysis of the arms in syringomyelia is not associated with increased tendon reflexes in the arms, whilst this is usually the case in amyotrophic lateral sclerosis; moreover, the latter disease runs a much more rapid course than syringomyelia.

Syringomyelia may cause spastic spinal paralysis similar to that in "primary lateral sclerosis," but in the latter affection the sensory disturbances of syringomyelia and muscular atrophy are absent.

Intra-medullary tumours of the spinal cord occasionally simulate syringomyelia, but the development of symptoms is much more rapid in the former disease.

Syringomyelia is distinguished from cervical pachymeningitis by the more rapid course of the latter; also in cervical pachymeningitis pain in the neck is a more prominent symptom, and when anæsthesia is present all forms of sensation are affected.

Spinal hæmorrhage occasionally simulates syringomyelia, and in both diseases dissociated anæsthesia may be present. But in spinal hæmorrhage the very sudden onset of the symptoms, and the tendency for gradual improvement in course of time, are points of great diagnostic importance, since in syringomyelia the onset of symptoms is very gradual, and the tendency is for slow and steady increase in their severity. In hysteria occasionally sensory symptoms occur which simulate those of syringomyelia, and in true syringomyelia hysterical

symptoms may be present. But in hysteria the localised muscular atrophy of the hands, spastic paresis and spinal curvature are absent, and the sensory symptoms may be influenced by mental conditions.

From tabes the diagnosis is usually easy. But in doubtful cases the Argyll-Robertson's pupil, the paralysis of ocular muscles and optic atrophy would be in favour of tabes. The onset of the disease before the age of twenty-five in an individual who had never suffered from syphilis would be in favour of syringomyelia.

Caries of the cervical vertebræ may cause atrophy of the hand muscles and spastic paralysis of the legs with sensory disturbances, and thus cause symptoms which might be mistaken on superficial examination for syringomyelia. But in cervical caries there are not the peculiar dissociated sensory symptoms (loss of sensation to pain and temperature whilst tactile sensation is preserved). Also the bone symptoms of caries, and signs of tubercular disease elsewhere, usually render the diagnosis easy.

Prognosis.—The disease is progressive, and, as regards recovery, the prognosis is very unfavourable. The patient is, however, often able to follow his employment for a long time. Also the disease in rare cases appears to become arrested, and remissions are not infrequently noted.

Treatment.—Unfortunately there is no treatment known which will arrest the disease. The treatment for the spastic condition of the legs and for bladder troubles described in the account of myelitis would be suitable in cases of syringomyelia. If pain is troublesome, the remedies may be employed which are recommended for the pain of tabes (especially cocaine).

Over-strain of the arms should be avoided, and the patient should be warned of the danger of injury to the limbs through the loss of sensation for pain and heat.

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Peroneal Type of Muscular Atrophy.

(Form described by Charcot, Marie and Tooth.)

This is a chronic progressive form of muscular atrophy of nerve origin, which is often hereditary. Frequently several members of a family suffer, and the disease may be traced through four or five generations. In some instances, however, brothers and sisters are affected, and there is no history of the disease amongst other relatives. In other instances no family history of the disease can be obtained.

Males are more frequently affected than females (5 to 1,—Sainton).

The affection usually begins in childhood, but sometimes later in life. (Onset from second to fortieth year, but mostly under the age of twenty-two.)

Symptoms.—The onset of the disease is gradual; the symptoms are almost always symmetrical, and the parts of the limbs most distant from the trunk are first affected. Usually the feet and lower part of the legs are first affected, and a long period (often several years) elapses before the arms suffer. Sometimes the arms are affected first.

When the disease begins in the usual manner, the small muscles of the feet suffer at an early period; they become wasted, but the changes at this region are apt to be overlooked until other muscles suffer. The muscles next affected (or affected at the same time as the foot muscles) are the peronei, the extensor longus digitorum, the extensor longus pollicis and the tibialis anticus. These muscles waste, and then attention is attracted to the disease. The gait is affected; the feet and toes are dropped, and the gait has the high stepping character. In course of time the feet become deformed and pes equinus or varo equinus develops. The toes become claw-like, the terminal phalanges being flexed and the proximal hyper-extended. In time the calf muscles undergo atrophy, and the whole leg becomes wasted. Later, atrophy extends to the thigh muscles. The atrophy is associated with weakness (paresis), and the two symptoms advance together.

When the arms become affected, the small muscles of the hand (thenar, hypothenar and interossei) gradually waste as in progressive muscular atrophy, and in time the claw-shaped hand is produced. Then the forearm muscles undergo atrophy: the extensors suffer before the flexors. After a long period the upper arm and shoulder muscles become atrophied. Hanel reports cases in four generations in which only the arm muscles were affected.

The muscles of the trunk remain unaffected longest, and in most cases they do not suffer.

The face muscles are usually not invaded, but in old standing cases they may be affected finally.

Bulbar symptoms do not occur.

Fibrillary twitchings are seen in the muscles; usually they are slight, but sometimes well marked.

On electrical examination the excitability of the nerves (going to the affected parts) is lost or diminished, both to galvanism and faradism; the muscles sometimes present simply diminished excitability to galvanism and faradism; in some cases there is the partial, in other cases the complete, reaction of degeneration. Schultze states that the complete reaction of degeneration may be sometimes detected at an early period. Finally the muscles may cease to react either to faradism or galvanism. It has been pointed out that electrical changes may be

detected in the muscles, before the atrophy or weakness. The mechanical excitability of the muscles is diminished.

The knee-jerks and deep reflexes are *absent* or greatly diminished; they are never increased.

Often sensation is not normal, but there are never marked sensory changes. Paræsthesia and painful sensations in the limbs are common, and sometimes there is slight but distinct diminution of cutaneous sensibility. Occasionally pain is severe in the limbs, but the nerve trunks are not painful. The bladder and rectum are not affected.

The course is very chronic; remissions may occur, or the disease may become arrested. The duration is many years. Death occurs from some intercurrent disease.

Pathological Anatomy.—The disease is regarded by many observers as a chronic multiple neuritis. In favour of this view are the slight sensory symptoms and pains (which are often present), the electrical changes, and the results of pathological examination.

By others the disease is regarded as a spinal neuritic muscular atrophy, or a degeneration of the peripheral motor and sensory neurons. In the muscles atrophied fibres are numerous, and in part they are replaced by connective tissue and fat.

In the peripheral nerves the intra-muscular fibres have been found markedly degenerated, whilst in the larger nerve trunks the nerve fibres are much less affected. The peripheral motor nerves are most affected, and the changes consist in a progressive degeneration extending from the peripheral towards the central part. At a late period the sensory peripheral nerves are also affected. The fibres of the anterior and posterior nerve roots have been markedly affected in some cases, but only slightly degenerated in others.

The nerve cells in the anterior horns of grey matter have presented little or no change in many cases; in others they have been markedly atrophied and degenerated, and similar changes have been found in the nerve cells of Clarke's columns.

Degeneration (sclerosis) has been found in the posterior columns, especially in Burdach's, and to a slight degree in Goll's columns. Also a slight degeneration has been found in the crossed pyramidal tracts.

Sainton concludes, from a careful study of the microscopical examination of the nervous system, that the pathological basis of the disease is:—

1. A sclerosis of the columns of Goll and Burdach;
2. Probably an atrophic lesion of the cells of the anterior horns;
3. Changes in the peripheral nerves more or less intense, sometimes minimal.

The **diagnosis** is usually easy. In idiopathic muscular atrophy (muscular dystrophy) there is no reaction of degeneration on electrical examination; sensory troubles are absent; the affection begins in the trunk muscles, or in the muscles of the limbs close to the trunk.

In progressive muscular atrophy and amyotrophic lateral sclerosis sensory symptoms are also absent. Peripheral neuritis of the usual form has a more rapid course, and after the symptoms have been present for many months, or a year or longer, there is no increase of the paralysis and wasting, whilst in the peroneal type of muscular atrophy the disease is steadily progressive.

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Hereditary Progressive Spinal Muscular Atrophy in Childhood.

Cases of a rare form of spinal muscular atrophy beginning in early childhood have been described by Werding, Hoffmann, Thompson and Bruce, and others.

This affection commences in the first year of life, in children who appear otherwise healthy. The first symptom is symmetrical flaccid atrophic paralysis of the muscles of the thigh and pelvis. The paralysis then extends to the muscles of the back, abdomen, neck, and shoulder girdle, afterwards it invades the muscles of the arms and legs in a descending order from the trunk towards the distal extremities. The knee-jerks are lost and the muscles give the reaction of degeneration. Frequently obesity is present. Sensory disturbances are absent. Contractions may develop later. The sphincters are not affected. The mental condition and general development are good ; the special senses are normal. Bulbar symptoms were absent in the cases recorded by Hoffmann, but present in one of Werding's cases. Muscular hypertrophy and pseudo-hypertrophy are absent. Death occurs in from one to four years after the onset of the disease, usually through secondary lung affections. More than one child of the family may be affected.

The diagnosis is easy. The symptoms—flaccid paralysis, atrophy, loss of knee-jerks, reaction of degeneration, with normal sensation and absence of pseudo-hypertrophy—indicate a neuropathic and especially a myelopathic origin of the disease. The peroneal type of muscular atrophy begins in distal extremities of the limbs, and advances towards the trunk (ascending course) ; and subjective sensory disturbances are usually present, often with objective disturbances. The gradual onset, progressive course, and fatal termination distinguish the disease from acute anterior poliomyelitis of the infant.

In three cases examined by Hoffmann symmetrical well-marked degeneration of the peripheral neurons of all the motor nerves below the hypoglossal nerve was found. The spinal accessory motor neuron was also degenerated. Atrophy and marked degeneration of the ganglion cells of the anterior horns of the spinal cord were found and almost complete atrophy of the intra- and extra-medullary anterior roots. Degeneration of the motor peripheral nerves and of the intra-

muscular branches was detected. There was also simple atrophy of muscle fibres up to complete disappearance · sometimes fatty degeneration of the fibres was present, sometimes absent. The posterior nerve roots were not affected. The white matter of the cord was normal, with the exception of the pyramidal tracts, which usually showed only slight changes.

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SECTION IX

DISEASES CAUSING SPASTIC PARESIS AND PARALYSIS

We have considered in the preceding chapters the chief diseases which give rise to paralysis with wasting. There is another important group of spinal diseases, in which a prominent symptom is paralysis, or paresis with rigidity of the legs—spastic paralysis. Both legs are usually affected, and hence the condition is described clinically as spastic paraplegia. This condition is associated with degeneration in the crossed pyramidal tracts of the spinal cord.

Spastic paraplegia is a clinical term, and not the name of a disease. It is the term applied to a group of symptoms: these symptoms occur in a number of spinal affections.

The condition known as spastic paraplegia is characterised by :—

1. Loss of power in both legs, with or without affection of the arms.
2. Rigidity of the affected limbs.
3. Increase of the deep reflexes, with the extensor type of plantar reflex and ankle-clonus.

Spastic paraplegia is usually secondary to some other spinal disease, or is associated with other symptoms indicating that the lesion of the cord is not limited to the crossed pyramidal tracts. But in a small group of cases the disease is probably primary. In this last group of cases there are simply the symptoms of spastic paraplegia, and no indication of lesion of any part of the cord except the crossed pyramidal tracts, and this diagnosis is confirmed pathologically.

The following are the diseases in which spastic paraplegia occurs :—

AFFECTIONS OF THE NERVOUS SYSTEM CAUSING SPASTIC PARAPLEGIA.

A. Spastic Paraplegia secondary to other Spinal Diseases.	Compression myelitis from— Vertebral caries. Vertebral tumour. Tumour of the spinal cord and its membranes. Spinal hydatid. Spinal compression from aneurism. Fracture-dislocation. Gunshot and other wounds of the cord. Acute transverse myelitis and disseminated myelitis. Hæmatomyelia and meningeal hæmorrhage. Spinal meningitis and pachymeningitis. Syphilitic myelitis, meningo-myelitis.

B. Spastic Paraplegia usually associated with other Symptoms.	Amyotrophic lateral sclerosis. Disseminated sclerosis. Syringomyelia. Ataxic paraplegia. Combined postero-lateral degeneration and sclerosis, associated with anæmia and various toxic conditions. Erb's syphilitic spinal paralysis. Occasionally general paralysis of the insane. Cerebral diplegia (Little's disease).
C. Primary Spastic Paraplegia.	Primary lateral sclerosis, and hereditary spastic paraplegia.

We have seen that in poliomyelitis, and in the spinal form of progressive muscular atrophy (Duchenne-Aran form), the disease affects only the lower motor neurons (*see* Fig. 87). In amyotrophic lateral sclerosis both upper and lower motor neurons are affected. In the small group of cases described as primary lateral sclerosis the degeneration is believed to affect only the upper motor neurons (*see* Fig. 120).

PRIMARY LATERAL SCLEROSIS

(Gr. σκληρός hard.)

(*Primary spastic paraplegia.*)

Clinically a condition is occasionally met with, in which there is the same spastic condition of the legs that often develops after many transverse lesions of the dorsal region of the spinal cord; but there are no sensory symptoms, no affection of bladder or rectum, no atrophy of the arm muscles, and no indications that any part of the cord is affected except the lateral pyramidal tracts. Erb in 1875, and Charcot soon afterwards, drew attention to these cases. Erb gave to the disease the name of primary lateral sclerosis. Charcot had previously shown that in amyotrophic lateral sclerosis muscular rigidity was associated with degeneration of the lateral pyramidal tracts; and both Erb and Charcot suggested, that in the cases of apparent primary spastic paraplegia the lesion was a degeneration of crossed pyramidal tracts, without affection of other parts of the cord. They suggested that there was a primary disease of these tracts, i.e. a primary lateral sclerosis.

This view has been disputed by many neurologists. Most of the cases regarded as primary lateral sclerosis during life have been found to be due to some other affection on pathological examination of the cord—very often to disseminated sclerosis, spinal syphilis, postero-lateral sclerosis etc. Even Erb admits now that primary lateral sclerosis is a much rarer affection than was formerly supposed; but he has collected (1902) ten cases, recorded comparatively recently, in which he believes that the post-mortem examination of the cord and the clinical course of the disease justify the view that primary lateral sclerosis does occur.

In all of these ten cases there was a pure lesion of the pyramidal

tracts, but, in addition, slight lesion of the direct cerebellar tracts, and trifling sclerosis of the tracts of Goll. The changes in the direct cerebellar tracts and the columns of Goll are of secondary importance; they had caused no clinical symptoms, and occur in many affections under the most varied circumstances. As Erb points out, we still regard tabes as a sclerosis of the posterior tracts, although changes are also sometimes found in the lateral and anterior tracts or in the grey matter. Moreover, it is important to remember that in the case of primary spastic paraplegia the post-mortem examination is only made, as a rule, after the disease has been present for very many years. Erb considers, therefore, that the pathological basis of primary lateral sclerosis is firmly established.

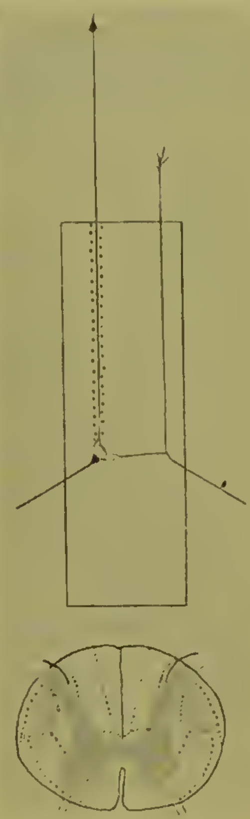

Fig. 120. — Primary Lateral Sclerosis. Lower figure = transverse section of Cord. Shaded part (in lateral pyramidal tract) indicates the position of the lesion. The upper figure = longitudinal section of Spinal Cord. To the left the anterior nerve root and its cell (lower motor neuron) is indicated. Above it is the upper motor neuron. To the right is the sensory neuron, posterior root and its collateral. Note degeneration affects the neuron marked with dots.

Microscopical examination shows that the grey substance, the pia mater and the vessels are normal. The degenerated lateral tracts present simple atrophy of the nerve fibres, with sclerosis of the neuroglia, but without vascular changes.

Symptoms.—The onset of the disease is very gradual. The patient notices a sensation of heaviness and dragging of the legs when walking; he soon becomes tired; and the legs become weak and stiff. There is slowly increasing difficulty in walking, and the gait becomes more and more laboured and dragging. But there is no pain, or only the pain of fatigue after exertion. Paræsthesia is absent, or if present it is only slight. When the affection is fully developed examination reveals only three groups of symptoms: (1) marked rigidity of the legs; (2) increased reflexes; (3) weakness of the legs, with impairment of the movements.

On attempting to make passive movements of the legs stiffness or resistance is experienced. Often this resistance is very great, and in a well marked case there is strong extensor spasm when the legs are extended. Often there is a condition which has been described as the "clasp knife rigidity"; the legs are exceedingly stiff when they are in the extended position, and on attempting to make passive flexion at

the knee joint the greatest difficulty is experienced at first; but when once the knee has been slightly flexed, when this initial resistance has been overcome, then the rigidity diminishes greatly, and the knee can be bent easily. (Just as occurs in the closing of a clasp knife, the resistance is greatest at first, and then rapidly diminishes.) The feet are often in a position of plantar flexion, owing to contraction of the calf muscles, and it is very difficult or impossible to dorsi-flex them, either voluntarily or passively. During a warm bath the rigidity is much less. The rigidity and loss of power are proportionate.

The gait is spastic. The steps are short, and the movements of the legs restricted. The legs are not raised high enough, and not put forward to the normal extent. They are drawn forwards as rigid bars; the knees are kept extended; the advancing leg is swung round in a semicircle; the toes scrape the ground; the feet are scarcely raised; and the gait is shuffling. The patient has a tendency to walk on the toes and the boot toes are worn down. Often there is marked spasm of the adductors of the thigh during walking, and a tendency for the advancing leg to be crossed in front of the stationary leg. Walking becomes more and more difficult, until the patient is unable to get about without the aid of a stick. Often the big toe of the advancing leg catches the heel of the stationary leg, and the patient falls to the ground.

When the rigidity is great if one leg be raised in the extended position, the other leg and the pelvis follow the movements. Finally, at a late period of the disease, the whole trunk follows the passive movements of the legs; and the same occurs also on passive abduction of one leg. In rare cases, at a late period the legs become flexed at the hips and knees.

The arms are often unaffected; but in some cases they are weak and rigid, and flexed at the elbow and wrist, as in cases of old hemiplegia.

There is marked increase of the knee-jerks and tendo Achillis reflexes; ankle-clonus is present on both sides; and there is often involuntary clonus of the foot when the patient is sitting with the feet in certain positions. A rectus-clonus can be obtained by suddenly depressing the patella (the leg being extended). The deep reflexes of the arms may be also increased.

The skin reflexes are usually increased. Babinski's extensor type of the plantar reflex is obtained in many cases.

When the leg is drawn up to the body (trunk), a powerful contraction of the tibialis anticus occurs, with marked prominence of its tendon. This associated movement is described as the tibialis phenomenon (Strümpell).

The muscles of the spastic limbs do not undergo atrophy; there are no fibrillary contractions; there is no reaction of degeneration; and the electrical reactions are usually normal. There are no trophic symptoms. The bladder and rectum are unaffected. There is no ataxia; no Romberg's symptom; no affection of the generative organs.

Sensory symptoms are usually entirely absent; occasionally there is slight numbness and tingling in the limbs, and occasionally slight dull pain; but there is no anæsthesia, and otherwise sensation is unaffected.

There is no affection of speech, of the cranial nerves, or of the sense organs. The pupils are not altered and memory is unimpaired.

Strümpell has described several forms of the disease.

1. In one form the symptoms correspond to those just described: there is no hereditary tendency; no symptoms of bulbar paralysis develop, and there is no atrophy of the small muscles of the hands, at least for a long period. But after many years a slight atrophy of the hand muscles may develop. This atrophy, however, usually remains very slight. (Various transitional forms may, however, occur between lateral sclerosis and amyotrophic lateral sclerosis.) In this form of lateral sclerosis involuntary attacks of laughing and crying often occur. The disease usually begins in advanced life. The slight affection of the hand muscles indicates that the degeneration has extended finally to the lower motor neuron—to the nerve cells of the anterior horn.

Pathological examination reveals sclerosis of the crossed pyramidal tracts extending up to the internal capsule. The nerve cells of the anterior horns are normal; or they present commencing degeneration, when there is slight affection of the small muscles of the hands. The columns of Goll and the direct cerebellar tract are not affected.

2. In a second form several members of the same family suffer. Strümpell describes it as the hereditary or family form of spastic spinal paralysis (endogenous form). Males are affected more frequently than females). The disease begins between the ages of twenty and thirty. It advances very slowly, and the duration is very long (twenty to thirty years). The legs gradually become rigid, and the tendon reflexes increased. The gait becomes spastic. The spastic symptoms are more marked than the paresis. Marked paresis is absent for a long period, and it is only very gradually that the muscular power diminishes. The arms and cerebral nerves are unaffected. The post-mortem examination reveals a pure sclerosis of the crossed pyramidal tracts, which, as a rule, extends to the cervical region. But finally there may be slight degeneration in the columns of Goll, in the cervical region, and sometimes also degeneration in the direct cerebellar tracts.

Newmark records a family of fourteen children, of whom six suffered from the disease; and another family of four children, of whom two suffered. E. Jones has recorded a family of nine children, of whom eight suffered from the disease.

3. A third form is the family infantile spastic spinal paralysis. The disease begins from the age of three to six years, or later. The symptoms correspond to those of spastic paralysis of the adult. The arms are sometimes unaffected or almost unaffected; but in other cases they are implicated. Often, though not always, there is a defective mental

condition. The exact nature of these cases can only be decided, when a greater number of clinical and pathological records have accumulated.

The course of primary lateral sclerosis is usually exceedingly chronic. In the only case in which I have ventured to give a definite diagnosis of primary lateral sclerosis the symptoms were of twenty years' duration, and during this period there had been nothing to indicate that any part of the nervous system had been affected except the crossed pyramidal tracts. Erb mentions a case in which the symptoms had been present twenty-six years. In rare cases the disease runs a more rapid course. Death occurs from marasmus or intercurrent diseases.

Diagnosis.—The diagnosis of primary lateral sclerosis can only be made when all of the other affections in which spastic paraplegia occurs can be excluded. These affections are enumerated on p. 263. Usually when a diagnosis of primary lateral sclerosis has been made, the development of additional symptoms, at a later period, shows that the patient is suffering from some other disease.

Hence the diagnosis of primary lateral sclerosis cannot be regarded as at all satisfactory unless the case has been followed for several years and occasionally even in the last few weeks of life symptoms appear for the first time, which show that the disease is not primary lateral sclerosis. Further, the post-mortem examination often shows that a case which has been regarded as one of primary lateral sclerosis up to its fatal termination has been due to some other affection.

A diagnosis of primary lateral sclerosis should not be maintained clinically, if any of the following symptoms are present:—Anæsthesia, ataxia, localised atrophy of muscles, affection of the bladder or rectum, tremor, pupillary or ocular changes. The clinical features must be limited to the three groups of symptoms already described—weakness of limbs, rigidity, and changes in the reflexes.

The following diseases causing spastic paraplegia have been most frequently mistaken for primary lateral sclerosis—disseminated sclerosis, amyotrophic lateral sclerosis, ataxic paraplegia, and various forms of combined postero-lateral sclerosis.

The typical forms of disseminated sclerosis are distinguished from primary lateral sclerosis by the presence of nystagmus, scanning speech and intention tremor. Optic atrophy, if present, would be in favour of disseminated sclerosis, and would exclude primary lateral sclerosis. The difficulty occurs chiefly in the anomalous cases of disseminated sclerosis, in which nystagmus, scanning speech, and tremor are all absent, and the chief symptoms are those of spastic paraplegia. In such cases it is only by repeated examination, and by watching the case for a long period, that the diagnosis can be satisfactorily made. The occurrence of any symptoms which cannot be included under the three groups just described (i.e. weakness, rigidity and exaggerated reflexes) definitely excludes primary lateral sclerosis. In two cases which the

writer had the opportunity of watching for long periods, the diagnosis of primary lateral sclerosis was finally excluded by the development of bladder symptoms during the last month of life. The microscopical examination of the nervous system showed, in both cases, that the disease was disseminated sclerosis.

In cases which otherwise present the symptomatology of primary lateral sclerosis, the presence of decided wasting of the small muscles of the hands would indicate amyotrophic lateral sclerosis ; the presence of ataxia would point to ataxic paraplegia or postero-lateral sclerosis.

In the diagnosis between hysterical rigidity of the legs and primary lateral sclerosis the "clasp knife rigidity" (rigidity which is most marked on first attempting to make passive flexion at the knee, and diminishes when a slight flexion has been produced) is in favour of the latter disease. Rigidity of this type does not occur in hysteria ; also *true* ankle-clonus probably never occurs in hysteria, though often a few jerks (clonus which is not sustained) may be obtained. Rectus-clonus occurs in primary lateral sclerosis, but not in hysteria. Babinski's extensor type of the plantar reflex occurs in many cases of primary lateral sclerosis, but not in hysteria. The presence of the Babinski's reflex is of diagnostic value, but its absence is not quite conclusive evidence, since it is occasionally absent or indefinite when the other symptoms of spastic paraplegia are present. The characteristic spastic gait does not occur in hysteria. Also in hysteria the onset of the spastic paraplegia is usually acute or rapid, and other hysterical symptoms are often present.

When spinal syphilis produces spastic paraplegia bladder symptoms or some form of anæsthesia or sensory affections are usually present.

In the congenital infantile spastic paraplegia of cerebral origin (one form of Little's disease) the diagnosis can be made by the history of the disease commencing at a very early period of life ; also there are often irregular movements in the fingers, and a slight degree of inco-ordination in the hands.

Great caution is therefore necessary in diagnosing primary lateral sclerosis. I believe, however, that a clinical diagnosis of primary lateral sclerosis is occasionally justifiable. In the case to which I have already referred, at the end of twenty years there were no symptoms except paresis of the legs, with great rigidity, and the changes in the reflexes. Hence a clinical diagnosis of primary lateral sclerosis appeared justifiable.

Etiology.—Strümpell thinks some cases of primary lateral sclerosis are of endogenous origin (hereditary), whilst others are of exogenous origin, as a result of an infectious or toxic condition. If the disease affects more than one member of a family, the endogenous origin is almost certain ; and probably many isolated cases belong to the family or endogenous type. The younger the patient at the onset of the disease the slower the progress, the more typical the disease, the less the

indications of external injurious influences, the greater the probability of an endogenous origin.

Of the cause of the exogenous cases little is known. Syphilis may cause a primary combined system disease of the lateral and posterior columns; possibly other toxic conditions may cause degeneration of the pyramidal tracts. The more definite the external factors in the etiology the greater the probability of the exogenous nature.

According to Gowers and J. Taylor "the most frequent apparent cause is concussion of the spine, such as a fall on the back."

Treatment.—Medical treatment has little influence on the course of the disease. It is important that the patient should avoid fatigue at the early period when he is still able to walk. Rubbing of the legs in an upward direction is to be recommended, but electrical treatment is probably injurious. Long continued warm baths tend to diminish the rigidity temporarily. Liquor arsenicalis is worthy of trial. Sir W. Gowers and J. Taylor mention a case in which recovery occurred after a course of Turkish baths with treatment by arsenic given internally.

If there should be the slightest suspicion of a previous syphilitic infection, anti-syphilitic treatment should be tried (*see* p. 399).

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DISSEMINATED SCLEROSIS

(Gr. σκληρός, hard.)

Etiology.—Disseminated sclerosis is not a common disease, even in hospital practice, but it appears to occur more frequently in England than in America. At the Manchester Royal Infirmary, during the ten years in which the writer was Medical Registrar the number of medical in-patients was 13,864, the number of cases of diseases of the nervous system 2,294, and of these 61 were cases of disseminated sclerosis, 118 cases of locomotor ataxia, 6 cases of paralysis agitans. At the Ancoats Hospital, Manchester, amongst 2,870 medical in-patients (of whom only a comparatively small proportion suffered from diseases of the nervous system) there were 10 of disseminated sclerosis.

The statistics given in a recent discussion on disseminated sclerosis at the New York Neurological Society indicate that disseminated sclerosis is much less frequent in the United States than in England. In Germany it appears to be rather a common nervous disease in country districts.

The age of the patient at the onset is usually between eighteen and thirty-five. The fact that the disease is one which generally commences in early adult life, and very rarely after the age of forty-five, is of some

diagnostic importance. Sometimes the disease begins before the age of twenty (ten–twenty). The youngest patient I have seen was eighteen at the onset of the disease. Rare cases of the disease in children are on record, and in a few of these the diagnosis has been verified pathologically. The two sexes are affected about equally. There is usually no family history of the disease; but there are recorded several rare instances of mother and child having suffered from the disease (one verified pathologically by Eichhorst), and of several members of one family being affected (E. S. Reynolds).

Little is known as to the etiology. Sometimes the disease follows an acute fever, such as typhoid fever, small-pox, scarlet fever, diphtheria, cholera. In two of my cases it directly followed an attack of influenza, and in one case acute rheumatism. Cases have also been recorded by others in which the disease followed these two febrile affections. It appears probable that febrile affections, parturition, and injuries either play some part as exciting causes of the disease, or that they render more prominent a previous slight or latent condition.

Symptoms resembling those of disseminated sclerosis have been recorded after malaria, but these cases usually show a tendency to improvement and recovery.

Oppenheim and others have recorded cases of the disease in persons who presented symptoms of chronic poisoning by zinc, tin, mercury, and carbon-monoxide. But evidence of chronic poisoning by these substances is certainly quite exceptional. The disease has no relation to syphilis.

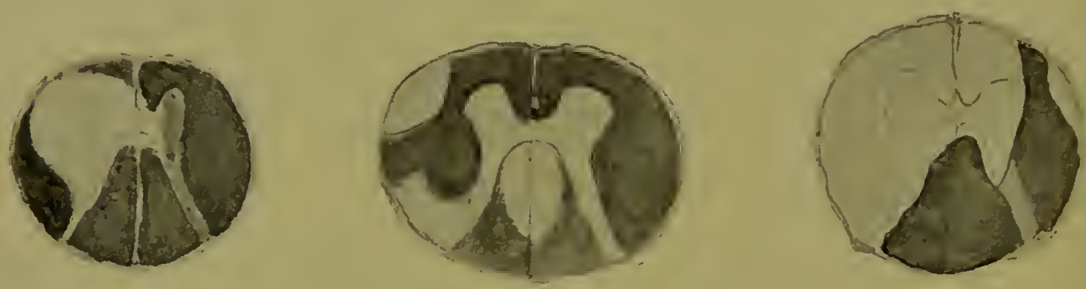
In the majority of cases of disseminated sclerosis the previous history presents nothing to which the disease can be attributed. (*See* Klausner's analysis of 126 cases.)

Strümpell regards the disease as one of endogenous nature, and attributes it to a congenital abnormality which leads to multiple proliferation of the neuroglia (multiple gliosis).

Pathological Anatomy.—The pathological examination of the nervous system in disseminated sclerosis reveals patches of disease—sclerosis—scattered about in the most irregular manner in the brain, pons, medulla, and spinal cord. The optic nerves and optic chiasma are often affected; the other cranial nerves rarely. Also the nerves of the cauda equina and the spinal nerve roots are occasionally found invaded by sclerosis. In the central nervous system both white and grey matter may be affected, though the former is perhaps more frequently the seat of disease. Parts of the nervous system adjacent to a diseased patch are normal. Usually both the brain and spinal cord present sclerosed patches (cerebro-spinal form); but sometimes they are found chiefly in the brain (cerebral form), in other cases chiefly in the spinal cord (spinal form). The patches are in some cases grey in colour and sharply defined; in others greyish white and less sharply defined. After hardening with Müller's fluid, the sclerosed patches are paler than the normal parts, and are seen better than in the fresh specimens. The smaller patches can only be recognised

by the microscope, but the larger are seen by the naked eye. Large patches in the cord, medulla and pons may extend over the greater part of the transverse section. The largest patches are seen in the white matter of the brain. The older patches are firmer than the normal substance of brain or cord ; but recent patches are occasionally seen, which are gelatinous and softer than the normal nervous tissue.

The chief macroscopic characters of the lesions in disseminated sclerosis are (1) their insular character ; (2) their irregular dissemination ; (3) the absence of secondary ascending and descending sclerosis in most cases. Even when almost the whole of the transverse section of the spinal cord is involved in the sclerosed patch, the posterior columns above the lesion, and the crossed pyramidal tracts below the lesion, usually show no signs of secondary degeneration. (*See* Fig. 121). This feature sharply distinguishes disseminated sclerosis of the spinal cord



a *b* *c*

Fig. 121.—Sections of Spinal Cord (Weigert's stain) in Disseminated Sclerosis. Pale areas = patches of sclerosis : dark portions = normal white matter : (*a*) upper cervical region ; (*b*) mid-cervical ; (*c*) lowest cervical. Note absence of ascending degeneration.

from other multiple lesions, such as disseminated myelitis and multiple syphilitic lesions, since in these two affections the spinal cord presents well-marked ascending and descending secondary degeneration (which can be seen by the naked eye in specimens hardened in Müller's fluid).

Under the microscope a striking feature of many patches is their *sharply defined nature*. The diseased patch ends abruptly, and the adjacent nerve tissue is quite normal. This feature is well seen in the sections stained according to Weigert's method. In other patches the margins are less sharply defined, and between the area of dense sclerosis and the normal tissue there is a zone in which slight changes are seen.

The patches of sclerosis are stained pale yellowish brown by Weigert's stain, whilst the adjacent healthy nerve tissue is black, owing to its being closely studded with normal nerve fibres, the medullary sheaths of which stain black in this method. In the sections stained with aniline blue black the sclerotic patches are stained a deeper colour than the normal tissue. Examination under a high power of the microscope shows that medullated nerve fibres are generally absent in the sclerotic patches ; but sometimes at the border of a patch there is a zone in which the medullated fibres are present, though scanty ; in other cases this intermediate

zone is practically absent, the disease patch ends abruptly, and the adjacent nervous tissue is quite normal. At the periphery of some patches of sclerosis compound granular cells are found, along with indications that the morbid process is still active. In the patches of sclerosis the axis-cylinders of the nerve fibres are very often present, though the medullary sheaths have disappeared (*see* Fig. 124). This is not invariably the case, but still the persistence of the axis-cylinders occurs much more frequently, and to a much greater extent, than in any other pathological condition. By the ordinary stains the axis-cylinders are not easily detected, but by the formol and nitrate of silver method, which stains axis-cylinders deep black (*see* p. 417), innumerable axis-cylinders can be seen in the sclerotic patches. In sections stained by this method diseased patches appear almost normal, whilst marked changes are seen in sections stained by Weigert's method or other stains. In the centre of a very old patch, however, the axis-cylinders may have degenerated as well as the medullary sheath, whilst the former are present in the more peripheral portion of the patch.

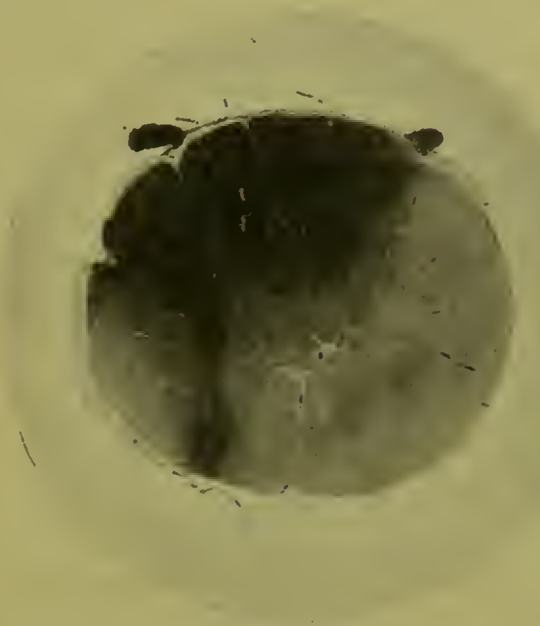


Fig. 122.—Photograph of Spinal Cord: Disseminated Sclerosis. Weigert's stain. Dark area normal; pale area degenerated. Note sharp margins of degenerated parts.

Longitudinal sections stained by formol and silver nitrate may show that the axis-cylinder is twisted or varicose, or it may present fusiform enlargements, and often the latter are fibrillated (Thomas).

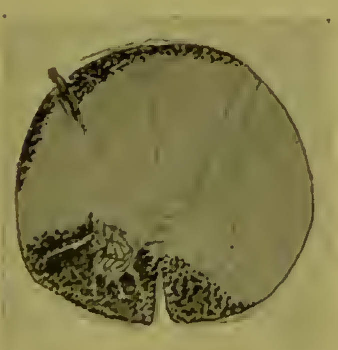


Fig. 123.—Spinal Cord: Disseminated Sclerosis. Pal's stain. Pale area degenerated; dark part normal.

The ganglion cells of the grey matter escape degeneration for a long period in a diseased patch; but at a very advanced stage they, like the axis-cylinders, may finally disappear. The fine nerve fibres of the grey matter degenerate or disappear, like those of the white substance.

The neuroglia connective tissue in a diseased patch is often greatly increased and converted into dense fibrous tissue. Spider cells may be present; but the neuroglia is not richer in nuclei than in the normal condition. In other cases the neuroglia,

though increased in amount, has an amorphous or homogeneous appearance under the microscope. In some patches the nerve fibres have degenerated, and spaces are left in the neurolgia from which the true nerve elements have disappeared ; but the connective tissue itself has not proliferated.

Compound granular cells are often present, especially at the periphery of a patch. They are most numerous in recent patches, but may be absent in old patches. Often there are large epithelial-like cells in cavities near blood-vessels.

FIG. 124.—Section of sclerotic patch in disseminated sclerosis (high power), Weigert's method. Deep black circles at upper part indicate white substance of Schwann (stained black) in sections of normal nerve fibres. The light-shaded area = sclerosed tissue. The white circles = spaces from which the nerve fibres have disappeared with the exception of the axis-cylinder. The faint dots in the shaded areas and in the white circles indicate axis-cylinders which have persisted.

The walls of the blood-vessels in some cases appear normal ; in some cases they are thickened and hyaline or present evidences of endo- or peri-arteritis ; in other cases (in recent patches) the peri-vascular lymph sheaths are filled with round cells, compound granular cells and fat globules. Often a large altered blood-vessel is found in the centre of a patch of sclerosis.

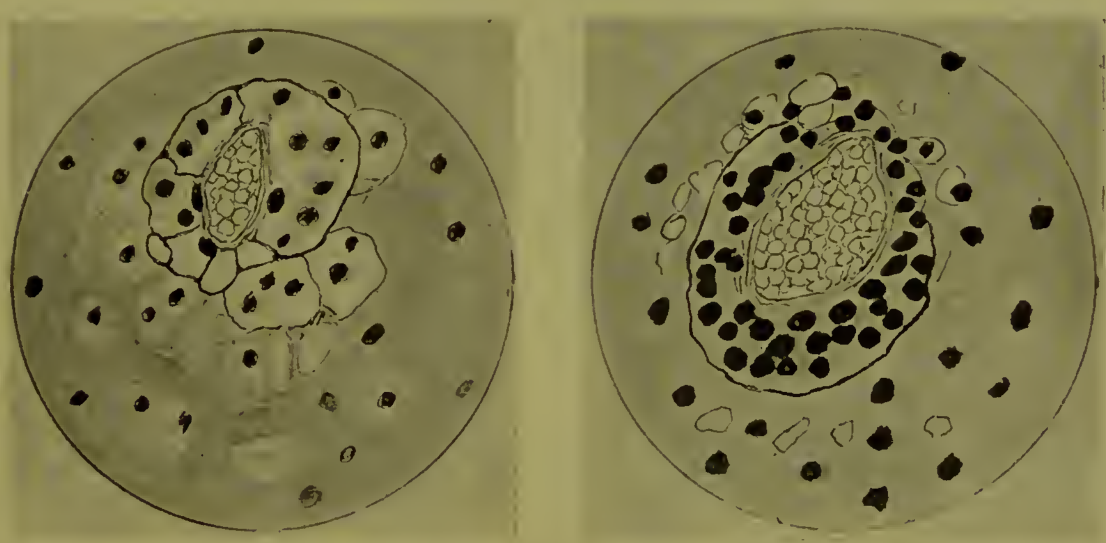
Four forms of sclerosed patches may therefore be met with—(1) patches in which nerve fibres have degenerated, leaving sieve-like small cavities in the neuroglia, whilst the neuroglia connective tissue itself has increased very little ; (2) patches in which there is marked proliferation of the neuroglia along with degeneration of nerve fibres ; (3) patches presenting diffuse proliferation of neuroglia, whilst the nerve fibres and cells persist ; (4) recent patches presenting changes very similar to those of cerebral softening—products of nerve degeneration and myelin drops, fat granular cells, and distension of the peri-vascular sheaths of the blood-vessels with round cells.

The absence of secondary degeneration is probably due to the persistence of axis-cylinders.

A point of importance is the presence of patches in various stages of development, in the same case ; some patches presenting firm sclerosis, whilst others are soft and recent, and may resemble ordinary softening. This fact shows, that some morbid condition or influence persists, which

continues to cause the development of new patches long after the onset of the disease.

Pathogenesis.—The exact cause of the morbid changes is still much disputed. By some observers the proliferation of neuroglia has been regarded as the primary change, and the degeneration of nerve fibres secondary. But others (including many recent observers) believe the degeneration of nerve fibres to be primary and the proliferation of neuroglia secondary; they believe the neuroglia proliferation to be caused by the degeneration of nerve fibres. In favour of the latter view is the fact, that in some cases there are diseased patches, in which the nerve fibres have degenerated, whilst proliferation of the neuroglia is slight or absent; and along with these patches are others presenting marked proliferation of the neuroglia. In a case which I have reported some diseased patches



A B

Figs. 125a and b.—Sections of small Blood Vessels of Spinal Cord in a patch of disseminated sclerosis. Lymph sheaths distended and filled with round nucleated cells (logwood stain).

presented the appearances of sclerosis, with proliferation of the neuroglia; whilst in others the changes resembled those of softening without any increase of neuroglia.

The very irregular distribution of the sclerosed patches, without any relation to nerve tracts or fibres, is suggestive of a primary change either in the blood or in the blood-vessels or lymphatics. Often marked vascular or peri-vascular changes have been detected, especially when the pathological examination has been made at an early stage of the disease. Such changes have been infiltration of the peri-vascular sheath with round cells in the more recent patches, or sclerotic or hyaline thickening of the vessel walls in the older patches. Sometimes a vessel with altered walls is seen in the centre of a diseased patch, and occasionally the extent of the area of sclerosis corresponds roughly to the area of distribution of a blood-vessel. Thus in a case recorded by the writer the extent of a

diseased patch corresponded roughly with the distribution of the anterior median arteries of the spinal cord at one region. Another point of interest in this case was the fact that in some sections the vessels *in the anterior median fissure* were dilated and surrounded by numerous round cells and nuclei, *before* they entered the substance of the cord. This appeared to indicate that blood or vascular changes were primary as regards the patch of sclerosis just referred to. Also in some sections a thrombus was seen in the anterior median vein, just at the commencement of the anterior median fissure.

Many years ago, Ribbert described peripheral thrombi (of white blood corpuscles), which partially obliterated the lumen of an artery in a diseased patch, and around these vessels were collections of round cells. In a few other cases thrombosis of vessels has been described.

Vascular changes are, however, often absent; but possibly this is owing to the changes having subsided long before the case terminated fatally. Moreover, the extent of the disease often does not correspond exactly with the distribution of a blood-vessel. There is much to be said against the view of actual primary disease of the *blood-vessels*, but much in favour of disseminated sclerosis being due to the circulation of some irritating or *toxic* substance *in* the blood-vessels and lymphatics. The lymphatic spaces between the adventitia and media of the blood-vessels have been found more or less obliterated by firm thick fibrous tissue; in other cases they have been distended. The peri-vascular lymphatic sheaths are frequently much dilated, and contain compound granular cells and granular degeneration products. It has been suggested that the changes in the lymphatics have caused obstruction to the flow of lymph, and the sclerotic changes have been regarded as a result of this condition. The importance of the cell walls of the capillaries and the lymphatics of the central nervous system, with respect to lymph circulation, is worthy of consideration. The cells of the capillary walls act as true secreting cells, according to Heidenhain, and when stimulated there is an increased flow of lymph into the surrounding tissues. Certain substances, such as toxins produced by some micro-organisms, have the power of stimulating the endothelial cells of the blood-vessels. As S. Woodhead pointed out some years ago, the endothelial walls of the cerebro-spinal system of capillaries and lymphatics are extremely delicate and active; the lymph flow is great; and if poisons, the products of bacteria, or bacteria themselves, have any effect on the walls of capillary vessels we should expect to find vascular and peri-vascular changes in the nervous system. Further, it is to be remembered that the lymphatics of the central nervous system are peculiar—they ensheath the blood-vessels, i.e. are vascular and peri-vascular (*see* p. 27).

The pathological changes in disseminated sclerosis are very suggestive of the presence of some irritating substance in the blood, which stimulates the endothelium of the walls of the vessels and the walls of the peri-

vascular lymphatics, and causes an extravasation of toxic lymph in the surrounding nerve tissue, with consequent degeneration of the myelin sheaths of the nerve fibres. The presence of recent patches, along with old patches, at the autopsy, shows that the morbid agent persists in the organism, and is able to cause the development of new patches of the disease long after the onset of the affection.

The sclerotic patches are scattered about in the nervous system in the most irregular manner, as already described; they may affect either white or grey matter of the brain or cord, they may affect cranial nerves, spinal nerve roots, or even nerves of the cauda equina. They have no relation to nerve tracts in the cord, and though they may sometimes be localised to the exact distribution of a blood-vessel, this is only occasionally observed. It is an interesting fact that the patches of sclerosis often present a very abrupt, straight, or curved, regular border, and that this margin or outline of a patch passes through the structures of the cord or brain, regardless of fibres, tracts, nerve cell or vessels (*see* Figs. 122, 123 and 126). The straight regular margin presents a marked contrast to the outline of a patch of disseminated myelitis, which is often very irregular or indented, and follows the course of vessels in streaks. The shape of the patches and their regular margins in disseminated sclerosis are suggestive of the infiltration of the nerve structures with a fluid of destructive character, the margins of the patch being determined by *physical conditions.* If a spinal cord, which has been hardened in Müller's fluid, be touched on its external surface, or on the cut surface of a transverse section, with a minute drop of deeply coloured fluid, such as a strong solution of aniline blue black, the fluid diffuses itself into the cord substance, and a stained patch is formed with a fairly regular, curved, clean cut margin—a margin which passes through the various structures of the cord abruptly and indifferently, and thus resembles that of a patch of disseminated sclerosis. This fact is suggestive of the patches of disseminated sclerosis being caused by

FIG. 126.—Section of spinal cord. Disseminated sclerosis. Weigert's stain. Central canal nearly in centre of figure. To the left is the grey matter with fine nerve fibres. The anterior and median parts of the grey matter are shown in the figure. The very dark portion (around the grey matter) is the white substance in transverse section. The very pale part is a large patch of sclerosis which surrounds the central canal. Note the *abrupt termination* of this patch in a *straight margin,* running nearly vertically through the grey matter.

the infiltration of the nervous system with a toxic fluid, their shape and margin being determined by *physical conditions*.[1]

Several views respecting the pathology of disseminated sclerosis may be mentioned. Strümpell regards the disease as one of endogenous nature, and as due to a multiple gliosis caused by a congenital abnormality. Against this view is the fact that there is very rarely any hereditary tendency; temporary improvement frequently occurs; also in some patches of disease there is no excess of neuroglia, but simply degeneration of nerve fibres. Leyden regards the disease as a form of chronic myelitis. Dejerine and Thomas think that the etiology, development, and pathological anatomy indicate that the disease is a variety of myelitis—possibly a mild and attenuated form of disseminated myelitis. But the pathological changes in disseminated myelitis and disseminated sclerosis differ, in three respects at least—(1) secondary ascending and descending sclerosis is caused by the patches of disseminated myelitis, but not, as a rule, by those of disseminated sclerosis; (2) the axis-cylinders are destroyed in disseminated myelitis, whilst they are long spared in disseminated sclerosis, and only disappear finally at the centre of a patch; (3) the margin of a patch of disseminated myelitis is often irregular and indented, and often streaks of myelitis follow the blood-vessels, whilst in disseminated sclerosis the margins are usually more regular and sharply defined, curved or straight. There is one form of disseminated sclerosis—the *diffuse* form—which closely resembles disseminated myelitis. In this form the patches are less sharply defined, and the axis-cylinders are degenerated; probably this form is really of the nature of a chronic disseminated myelitis. Demange has described diffuse disseminated sclerosis of this form, in which the margin of the patches were not sharply defined, but presented radiating processes ("*sclérose non en plaques mais in taches*").

Though the exact cause of the changes is still obscure, there are three considerations which appear to be worthy of careful attention.

1. The possibility of the patches being caused by the infiltration of the tissues with toxic lymph passed through the walls of the peri-vascular lymphatics.

2. The probability of the shape and outline of the sclerosed patches being determined by *physical conditions*, since patches of similar form can be produced when stained fluid is allowed to infiltrate the cord from various points.

3. The probability of a primary altered (toxic) blood condition.

Symptoms.—Owing to the variable and irregular distribution of the sclerotic patches, the modes of onset of the symptoms differ greatly, and

[1] Alcohol and several other chemical fluids have the power of dissolving out the white substance of Schwann from the nerve fibres of specimens of the cord and brain kept in these fluids for long periods, whilst the axis-cylinders and other structure are left. Thus a microscopical appearance is produced on section which somewhat resembles that of one form of the sclerosed patches (1). This fact is of interest, though the disease has no relation to alcoholism.

there are many forms of the disease. But it will be most satisfactory to sketch first the characteristic cerebro-spinal form, and afterwards to mention the atypical forms.

The course of the disease, in its typical form, may be divided into three stages:—the early period, in which the symptoms vary much and are often indefinite; a second period, in which well marked symptoms have developed; and a final period, in which there is failure of organic functions, and often paralysis of the sphincters, cystitis, and bed-sores.

The onset is usually gradual; but occasionally rapid. The most common symptoms of which the patient first complains (excluding objective signs such as nystagmus, optic atrophy and objective sensory disturbances) are a feeling of weakness of one or more limbs, numbness and other forms of paræsthesia in the hands or legs, tremor of the arms, legs, or head, impairment of vision, and occasionally vertigo and other cerebral symptoms. Often the symptoms are unilateral at first (18 per cent); and not infrequently there are early subjective sensory disturbances, alone or combined with other symptoms (*see* analysis of 80 cases by A. Mackintosh).

Oppenheim and others have pointed out that visual failure or optic atrophy is sometimes the first sign, and may precede all other symptoms for many years.

Whatever the mode of onset, the symptoms of disseminated sclerosis advance, though there are often remissions or periods in which the condition remains stationary for months or years.

In course of time, in a typical case, the most prominent symptoms are—(1) tremor in the arms; (2) nystagmus; (3) alteration of speech, "scanning speech"; (4) weakness, with spastic condition of the legs.

The knee-jerks usually become exaggerated, and ankle-clonus and the extensor form of plantar reflex often develop. Visual impairment and optic atrophy are frequent symptoms. Apart from paræsthesia, objective sensory symptoms are rare. Finally affections of the sphincters, trophic disturbances, bed-sores and cystitis may develop.

Three symptoms of the disease are of special diagnostic importance—(1) tremor, (2) nystagmus and (3) scanning speech.

Tremor.—The tremor of disseminated sclerosis occurs only on voluntary movement. During repose there is no tremor; but on attempting to carry out any movement, such as touching the tip of the nose with the index finger, or on attempting to grasp any object, tremor occurs. Hence it is spoken of as intention tremor, or volitional tremor. In attempting to carry out one of the movements mentioned the arm jerks about in an irregular manner, first in one direction, then in another, until, with great effort, the object aimed at is reached. The tremor becomes quicker towards the end of the voluntary act. It consists of irregular oscillations or jerky movements of a coarse form, occurring at the rate of about five or six per second. But owing to the wide range of the movements the tremor appears more rapid. The tremor is clearly brought out by the well-known method of asking the patient to drink from a glass

full of water. He seizes the glass, moves it towards his lips, but in doing so the hand begins to shake; the tremor increases as the mouth is approached, and the contents of the glass are spilled, and often the glass is knocked against the teeth. As Marie points out, the tremor is not limited to the extremity of the limb, nor to the muscles of the forearm; it affects the muscles about the scapula markedly. The tremor is increased by mental excitement, attention and effort, and by the patient's endeavour to check it. In mild cases tremor is only observed at the end of a voluntary movement; in severe cases it begins directly the action is commenced, and increases as the movement continues. Often it spreads to

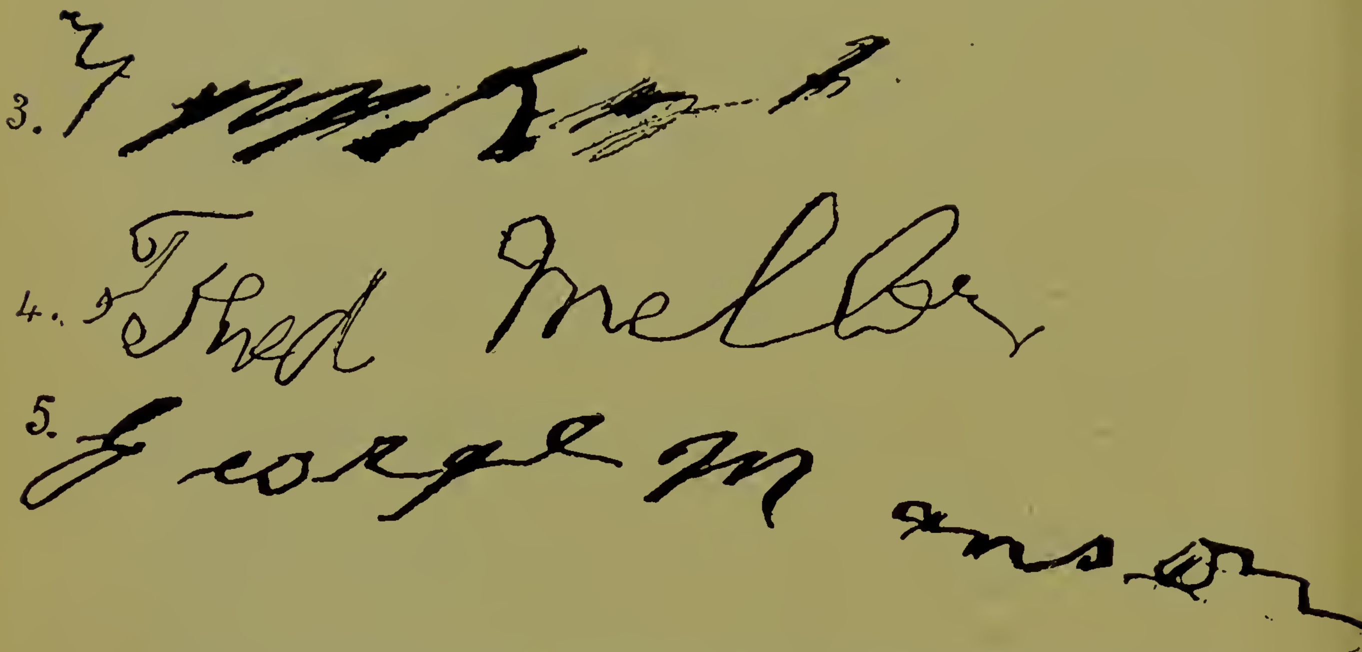


FIG. 127.—Handwriting: Disseminated Sclerosis. 3 and 4, advanced cases; 5, early case. Signatures of patients (Walker, Fred Mellor, and George Manson).

muscles not called into play in the movement. It occurs most frequently and is usually most marked in the arms; but the muscles of the head and neck and of the legs may also be affected. Sometimes the tremor is much more marked in the head and neck than in the arms. When the patient is sitting up in bed or on a chair, marked tremor of the head may be observed, even though he is not attempting to perform any movement of the head. This appears as if the tremor of the head differed from that of the arm, in occurring apart from voluntary movement. But observation will clearly show that when the head trembles, though the patient is not attempting to carry out any movement, still the muscles of the head and neck are in action in balancing or fixing the position of the head; and hence the tremor is one associated with contraction of these muscles. That this explanation is true is shown by the fact that the tremor is arrested when the head is allowed to rest on the pillow, so that the contraction of all muscles connected with it ceases.

The tremor of the arm interferes, of course, with all fine movement of

the limb. A female patient finds it difficult or impossible to thread a needle.

The handwriting is affected *early* and markedly. In severe forms, on attempting to use a pen, it is jerked so violently on the paper that only a few dashes are produced, and the pen is almost broken.

Fig. 127 (3) represents the result of the patient's attempt to write her name, in a severe form of the disease.

When the tremor is less severe the patient may be able to write; but the writing is jerky, the letters are very irregular, and this jerky irregularity is often best seen at the end of a word or sentence. There is a marked coarse irregularity in the general form of the letters, whilst the separate strokes produced by the pen often present no irregularities in

Fig. 128.—Lines drawn by patients suffering from Disseminated Sclerosis—P 3 and P 4. Lines drawn by the writer with same pen for comparison are marked W. W.

short portions. The movement of the pen is jerky, and often rapid towards the end of a word.

The handwriting may be markedly affected when the tremor of the arm is very slight (as shown in Fig. 127 (5)).

When the tremor of the hand is very slight the patient may be able to draw a very short line on paper fairly well; but if the tremor is well marked, the line shows very coarse irregularities (*see* Fig. 128).

Nystagmus or nystagmoid movements are present in half of the cases. The symptom is usually bilateral. In very marked cases, when the patient looks forward, lateral or vertical (occasionally rotatory) movements of the eyeball may occur. But in most cases it is only on looking to one side that rhythmical lateral movements of the eyes occur, to and fro, outwards and inwards (nystagmus). The movements are usually in the lateral direction; very rarely the nystagmus is vertical, and in exceptional cases it is rotatory. It is important to remember that on looking to one side a slight nystagmus (three or four oscillations) sometimes occurs in health, and therefore only a well-marked nystagmus is of diagnostic value.

Scanning speech.—The speech is often, but not invariably affected.

It is slow and deliberate; there is a tendency to divide the words into syllables; each syllable is pronounced separately, and there is an abnormally long pause between the syllables. The voice is monotonous and often husky. The speech is often like that of a child spelling the words slowly. The change is well brought out by asking the patient to quickly pronounce "Constantinople," "Manchester Royal Infirmary," "Lancashire and Yorkshire Railway Company."

Visual disturbances are present in a large number of cases, probably in about one-half of the cases. The visual defects in some cases develop gradually; in others acutely. Often after a time improvement occurs; but at a later period the sight fails again. Several relapses may occur. Sometimes the vision fails in one eye and then improves; afterwards

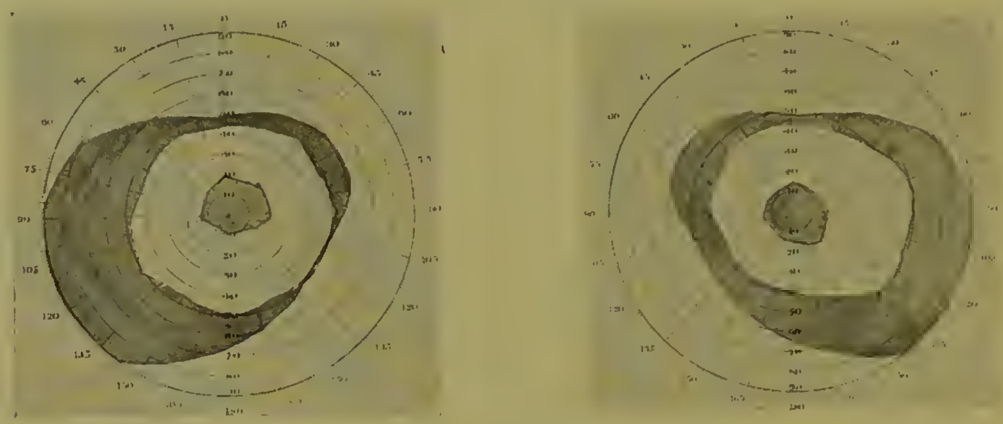


Fig. 129.—Fields of Vision: Disseminated Sclerosis, with visual failure. Central dark area = central scotoma.

visual failure may occur in the other eye, and this in time may subside. At the early stage there is sometimes complete blindness in one eye for a short period, and then the vision returns.

The visual defects scarcely ever proceed to complete permanent blindness. In some cases there is a central scotoma for white and colours, or for colours only, especially red and green (achromatopsy). In other cases there is irregular restriction of the field of vision. This restriction may be associated with a scotoma. Sometimes, especially at the early period, the visual defect is not associated with any change that can be recognized on ophthalmoscopic examination.

Occasionally marked improvement occurs and the vision may become almost normal. Both eyes are usually not affected equally. In many cases, but not in all, the visual defect is associated with ophthalmoscopic changes. These consist of partial optic atrophy. A portion of the disc, especially the temporal half, become very pale; sometimes the whole disc is very pale, or there is complete optic atrophy. (For excellent coloured illustration of this pallor of the temporal half of the optic disc see Strümpell's text-book of medicine, 1907, Fig. 66.) The ophthalmoscopic changes are usually much less marked than the visual defect. (Pathologically, patches of sclerosis are found in many cases in the optic chiasma

or optic nerve.) It is said that in rare instances the optic atrophy is preceded by slight neuritis, but real swelling of the discs rarely if ever occurs. Uhthoff found ophthalmoscopic changes in 45 per cent. of his cases (40 per cent. partial or marked optic atrophy; optic neuritis in 5 per cent.). Buzzard found pallor of the discs in 43 per cent.; he never detected a true optic neuritis, though he has seen a grey and hyperæmic appearance of the disc.

The optic discs are unequally affected or the affection is unilateral. Usually visual defect or optic atrophy is an early symptom. As already mentioned it may precede other symptoms for many years (Oppenheim).

Real *paralysis* is rare, except at the late period of the disease. For a long period all the coarse movements are performed without much diminution of power; but often the movements are slower, the patient is soon tired, and the same movement cannot be repeated so rapidly as in health. In course of time the patient feels a sense of stiffness in his movement, and rigidity may be detected on passive movements.

The paresis and rigidity may affect one limb only, or both legs, or arms and legs. At first it is often variable; in course of time spastic paralysis may develop, but the spastic condition is less marked in the arms than in the legs.

The deep reflexes, knee-jerks, triceps and wrist-jerks are increased. The ulnar and radial extensor jerks may be well marked on one or both sides (*see* p. 69). Ankle-clonus is often obtained as the disease advances. In exceptional cases the knee-jerks have been absent on one or both sides. The plantar reflexes are usually present, and very often of the extensor type (Babinski's reflex). Oppenheim's reflex is also often obtained. According to Strümpell the abdominal reflexes are so frequently lost that this symptom is of some diagnostic value. (In health these reflexes are occasionally absent, or very difficult to obtain. Strümpell states that they are lost in 13·5 per cent. of healthy individuals, and in 67 per cent. of the cases of disseminated sclerosis.)

Paralysis or paresis of the abdominal muscles may occur.

The gait at first is little altered; later it is frequently spastic; occasionally staggering or ataxic; or there is a combination of the ataxic and spastic gait. When the paraplegia becomes marked the patient is unable to walk.

Apoplectiform attacks occur in a small percentage of cases. The patient becomes unconscious; the face is flushed, and the temperature elevated. On recovery of consciousness hemiplegia is detected; but this rapidly disappears as a rule. Sometimes several attacks occur during the course of the disease.

Vertigo sometimes occurs, and it may be an early symptom. It may occur in paroxysms; it may occur when the patient looks upwards, or when he is walking.

Headache is an occasional symptom, but it is rarely severe. Marked

mental defects are very rare ; but the patient sometimes appears weak-minded ; his memory may fail, and he may take little interest in his surroundings. The patient is not infrequently very emotional ; curious rare symptoms are involuntary fits of laughter and crying, which occur without any cause, or after some very trivial cause. Often the fits of laughter are followed immediately by attacks of crying.

In very rare cases paralysis of the external ocular muscles has been observed ; in a few cases there has been associated paralysis of ocular muscles for lateral movement, whilst the power of convergence has been unaffected. A few cases of ophthalmoplegia are on record. Usually the pupils and pupillary reflexes are normal; immobility of the pupils is almost invariably absent. Occasionally inequality of the pupils or myosis has been recorded. In exceptional cases paralysis of the vocal cords has been noted, and occasionally tremor of the cords when the patient has pronounced the letter "e."

Marked sensory symptoms are exceptional. Often sensation appears practically normal, but after repeated examinations it may be found that slight sensory symptoms can be detected, though these are usually temporary. The chief sensory affection is temporary paræsthesia—numbness and tingling in the limbs. Occasionally there is diminution or loss of tactile or painful sensation, of the temperature sense, or of the sense of position. After a few days or weeks the sensory symptoms may disappear ; but sometimes they are permament. Pain in the limbs or joints is occasionally complained of, but it is seldom severe, and rarely, if ever, lancinating in character.

Marked or permanent bladder symptoms are rare, except at a late period ; but often slight and temporary affections are observed at an early stage—such as increased excitability of the bladder, slight difficulty in micturition, slight retention or incontinence. These symtoms are present for a few days or weeks, and then disappear, though they may return at a later stage. There may be complete loss of control of the bladder, usually at a late stage of the disease. Incontinence of fæces is very rare.

The electrical reactions are normal, and usually there is no atrophy of muscles. In exceptional cases atrophy of a group of muscles has been recorded, but usually in such cases there has been no reaction of degeneration on electrical examination.

Bed-sores may occur at a late stage of the disease ; usually they are associated with bladder symptoms.

Death occurs from emaciation, from cystitis, pyelo-nephritis and pyæmic conditions following paralysis of the bladder, from septic absorption from a bed-sore, or from some intercurrent disease, such as pneumonia or tuberculosis.

Forms of the Disease.—From the irregular and varied distribution of the lesions, it is not surprising that forms of the disease which differ from the typical variety already described are numerous.

Hysterical form. This is a form which causes great difficulty in the diagnosis, especially at the early stage. It has been carefully described by Buzzard. Women súffer more frequently than men, and the symptoms often develop after a great mental or physical shock.

The patient is emotional, and complains of numbness, deadness, or pins and needle sensation in some part of the body : or there is loss of power, sometimes with slight stiffness in one or two limbs. Often there are also distinct hysterical symptoms. Usually the loss of power is temporary ; recovery, partial or complete, occurs in course of time, and this is then regarded as a point in favour of the diagnosis of hysteria. But at a later date the symptoms return, or other symptoms appear, and the disease progresses, though one or more periods of remission of the symptoms may occur. Paraplegia may finally develop, with or without spastic condition. The knee-jerks become increased, and ankle-clonus and the extensor form of plantar reflex (Babinski's reflex) may appear. Impairment of vision is sometimes noted. Nystagmus, pallor of the optic discs, and tremor may finally appear ; but often these symptoms, as well as the scanning speech never develop. The mental condition remains unchanged, or the patient may become demented at a late period : the sphincters are sometimes affected, and cystitis and bedsores may develop.

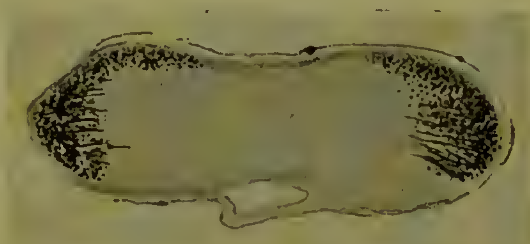


FIG. 130.—Transverse Section of Optic Chiasma (× 3 diameters), in case of disseminated sclerosis. Pale area = patch of sclerosis. Part marked with black dots and fibres = normal. Weigert's stain.

Spinal form. In this variety the symptoms are those of spinal spastic paraplegia, and death may occur without the three characteristic symptoms of the disease (nystagmus, tremor and scanning speech) or other cerebral symptoms having developed. In these cases the bladder often becomes paralysed shortly before death. In other cases the symptoms are those of spastic paraplegia with partial optic atrophy. In some cases the symptoms resemble those of a chronic transverse myelitis, and consist chiefly of paraplegia, paralysis of the bladder and sensory disturbances, whilst cerebral symptoms are absent or slight.

In another form *failure of vision* in one or both eyes is the first symptom of the disease, or the most prominent symptom at the onset. Often a long period—many months or many years—elapses before other symptoms develop, and hence these cases are frequently not diagnosed correctly at first. Sometimes there is improvement of vision, partial or almost complete in course of time ; but in other cases the visual failure becomes permanent and optic atrophy may develop. Ophthalmoscopic examination may reveal partial or marked optic atrophy, but in other cases the discs appear normal or almost normal (*see* p. 289).

Rare forms of the disease are met with presenting other peculiarities in the symptomatology. In a few cases bulbar symptoms have been prominent, but the affection has shown remarkable remissions. In rare instances the chief symptoms have been those of hemiplegia or hemiparesis of slow onset ; but the nature of the disease has often been indicated by the development of nystagmus, optic atrophy, tremor, etc.

Oppenheim has recorded cases of acute ataxia of the arms with bulbar symptoms, followed by typical symptoms of disseminated sclerosis. He believes that disseminated sclerosis may develop from a myelo-encephalitis which has followed an infectious disease.

So-called acute disseminated sclerosis. Recently attention has been drawn to a form of disseminated sclerosis which runs a very rapid course, death occurring within a short period, often in three or six months. The onset is gradual, the temperature is not elevated, and, though there are remissions, the affection is progressive. These cases have been described as acute disseminated sclerosis. Pathological examination shows degeneration of the medullary sheath of the nerve fibres, whilst the axis-cylinder is relatively intact. At the same time, or soon afterwards there is proliferation of the neuroglia. In a case which I have recorded, in a man aged twenty-three, death occurred within five months of the onset of decided symptoms ; though temporary slight and doubtful paræsthesia had been present longer, but not for more than twelve months. (In this case in addition to sclerotic patches in the cord and brain, there were others which, both macroscopically and microscopically, resembled those of softening.)

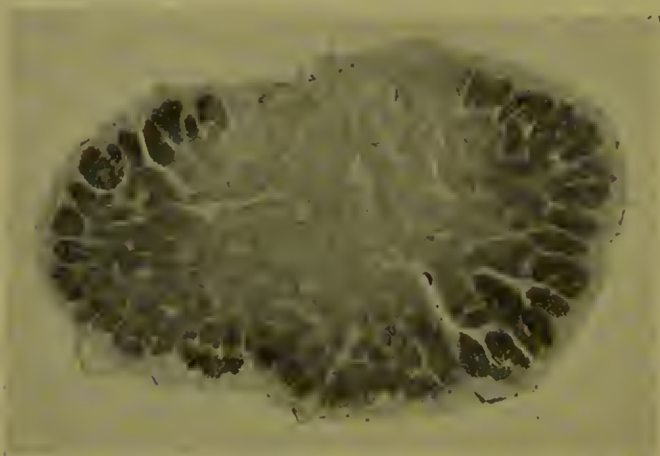


Fig. 131.—Microphotograph of transverse section of Optic Nerve: disseminated sclerosis. Weigert's stain. Optic atrophy and marked visual failure. Note patch of sclerosis (pale area) in centre of section.

Strümpell has described a *latent* form of disseminated sclerosis, in which the symptoms have consisted of slight headache and vertigo for a long period. Then temporary apoplectiform attacks, and later epileptic attacks have occurred. In such a case Strümpell found the changes of disseminated sclerosis post-mortem.

The **diagnosis** in a typical case is easy ; but it is important to remember that very many cases are atypical, and that the three characteristic symptoms (tremor, nystagmus, and scanning speech) as already mentioned, may appear very late, or may never develop at all. Probably the majority of cases do not present these three characteristic symptoms.

(It is estimated that scanning speech occurs only in 12 per cent. and intention tremor and nystagmus in 50 per cent. of the cases—Morawitz.)

The diagnosis from *hysteria* is often very difficult at the onset. "Of all organic diseases of the nervous system, disseminated sclerosis in its early stage is that which is most commonly mistaken for hysteria" (Buzzard). Hysterical symptoms are often present in disseminated sclerosis, and the diagnostic difficulty is increased by the great variability of the mode of onset of the disease, and by the fact that one or more periods of improvement or recovery may occur at the early stage. Hence in all cases of hysterical paralysis the diagnosis of disseminated sclerosis should be carefully considered. The following points, most of which have been specially emphasized by Buzzard, are of value in the differential diagnosis. The presence of the ophthalmoscopic signs of optic atrophy would be evidence in favour of disseminated sclerosis and would exclude hysteria. The presence of a central scotoma in the field of vision, or of nystagmus, would point to disseminated sclerosis : probably true nystagmus never occurs in hysteria. Babinski's extensor type of plantar reflex, if present, would be decidedly in favour of disseminated sclerosis : probably it never occurs in hysteria : in the latter affection the plantar reflexes are often absent. Loss of control of the sphincters is usually a late symptom in disseminated sclerosis and occurs long after the diagnosis of hysteria has been excluded : but if this symptom should occur at an early period it is important evidence against hysteria. The knee-jerks are usually present and increased in both diseases, absence of one or both knee-jerks is occasionally caused by disseminated sclerosis, but never by hysteria. In hysterical paraplegia the loss of power is usually complete, and the limbs are more often flaccid than rigid. In disseminated sclerosis complete paraplegia is rare except at an advanced stage. Also a spastic gait, marked and sustained ankle-clonus, and true intention tremor, occurring only on voluntary movement, are in favour of disseminated sclerosis. In hysteria there is often ataxia in the movement rather than tremor, and if the latter should be present it occurs when the arm is supported as well as during voluntary movement, and it often varies in its character. As Buzzard points out, the hysterical patient may be able to touch an object with the finger quite steadily, but when the finger has rested on the object for a few seconds tremor occurs, whilst in disseminated sclerosis the tremor is marked during the movement, but it tends to cease when the object is touched.

The spastic paraplegia occurring in disseminated sclerosis resembles that of *primary lateral sclerosis*. In many cases regarded as primary lateral sclerosis during life, the pathological examination has revealed disseminated sclerosis. In all cases diagnosed as primary lateral sclerosis the possibility of disseminated sclerosis should be considered carefully, since some of the atypical cases (spinal forms) of the latter disease strongly

simulate the former affection. Hence repeated examination should be made for nystagmus, intention tremor, scanning speech, optic atrophy with central scotomata, and affections of the bladder. These symptoms would be evidence in favour of disseminated sclerosis, and against primary lateral sclerosis. I have seen the spinal form of disseminated sclerosis run its course almost up to the fatal termination with symptoms of spastic paraplegia (lateral sclerosis); but paralysis of the bladder or marked nystagmus occurring during the last few weeks of life has shown that the affection had not been primary lateral sclerosis.

All the points just mentioned are also of importance in distinguishing disseminated sclerosis from *ataxic paraplegia.*

In rare cases there is difficulty in the differential diagnosis between disseminated sclerosis and *general paralysis* of the insane. But in the latter disease the tremor of the arms, when present, is more rapid and finer, and it occurs both on movement and when the limb is at rest. Also there is usually fine tremor of the lips and tongue. The speech in general paralysis is tremulous rather than slow and scanning. There is a tendency to repeat syllables, and the patient stumbles over syllables. In general paralysis mental symptoms occur early, whilst in disseminated sclerosis they only occur at a late period, and then are usually slight; also there are no grand ideas in disseminated sclerosis.

The Argyll-Robertson pupil is present in many cases of general paralysis of the insane, but does not occur in disseminated sclerosis.

From *tabes* the diagnosis is usually easy. The absent knee-jerks and shooting pains which are usual in tabes, are extremely rare symptoms in disseminated sclerosis. Also the nystagmus, scanning speech, intention tremor, and the Babinski plantar reflex are absent in tabes. The Argyll-Robertson pupil does not occur in disseminated sclerosis.

Freidreich's disease is usually distinguished from disseminated sclerosis by the absence of knee-jerks, the absence of spastic condition of the legs, the frequent presence of lateral spinal curvature and of pes cavus, and by the family history of the disease in the former affection.

Multiple cerebro-spinal syphilitic lesions may cause symptoms somewhat resembling those of disseminated sclerosis: but in the former affection nystagmus, scanning speech, and tremor are absent. In favour of multiple syphilitic lesion would be immobility of the pupil, marked optic neuritis, history of syphilis and good result of anti-syphilitic treatment. Symptoms of chronic meningitis do not occur in disseminated sclerosis, but are common in cerebral syphilis.

Tumours of the optic thalamus, crus and pons sometimes cause tremor resembling that of disseminated sclerosis. But these, and other cases of cerebral or cerebellar tumour can usually be distinguished from disseminated sclerosis by the presence of severe headache, vomiting and optic neuritis (with marked swelling of the disc) in the former affections. Severe headache and vomiting are unusual in disseminated sclerosis, and

in the very rare cases in which optic neuritis occurs the swelling of the disc is slight and the condition soon passes into atrophy. Usually there is *no neuritis* but *primary óptic atrophy* in disseminated sclerosis.

After malaria a pseudo-disseminated sclerosis occasionally occurs, but generally the symptoms subside under the use of quinine.

A disease of the motor cortex of both cerebral hemispheres in children, due to meningeal hæmorrhage or thrombosis of veins in the blood sinuses may cause symptoms somewhat similar to those of disseminated sclerosis—jerky inco-ordination of the arms and legs with affection of speech. But the history of these cases is sufficient for their recognition.

In *paralysis agitans* the tremor occurs during rest (i.e. when the arms are supported), whilst in most cases it ceases or diminishes during voluntary movement; also the tremor is finer, generally it is best marked in the hand between the fingers and thumb—producing the "pill-rolling" character of movement. The handwriting is little affected at the early period, the attitude is peculiar and stooping, the facial expression fixed, and propulsion or retro-pulsion may occur, whilst optic atrophy, ankle-clonus and the Babinski or extensor type of plantar reflex, nystagmus, and scanning speech are all absent. In disseminated sclerosis the disease usually begins before the age of forty; in paralysis agitans usually after that age. In disseminated sclerosis the handwriting is often affected markedly; and at an early stage of the disease the writing is often very irregular and difficult when the tremor of the arms is very slight.

Oppenheim, Müller and others have drawn attention to the importance of *optic atrophy and visual failure* in early disseminated sclerosis; as already mentioned they may be the first symptoms, or the only symptoms for many years. In a young person who has never suffered from syphilis, lead poisoning, or alcoholism, if amblyopia should suddenly develop in one eye and then partially subside, and if some time later a similar affection should develop in the other eye, the diagnosis of disseminated sclerosis is very probable (Müller). In such a case confirmatory signs would be pallor of the temporal half of the optic disc with central scotoma in the field of vision (as described on p. 282), loss of the abdominal reflexes, Babinski's extensor type of plantar reflex, and slight shaking of the hand in fine movements, as in writing. Another symptom which may occur early is an abnormal sensation of tiredness, often localised to the legs. The age of the patient twenty to forty, the absence of any recognisable cause, the absence of pain (in most cases), are also points in favour of the diagnosis of disseminated sclerosis.

Prognosis.—The course of the disease presents variations just as great as is shown by the symptomatology and the pathological distribution of the sclerosed patches.

The disease is usually of long duration and the termination is fatal, but death may not occur for many years, even ten or twenty or longer.

In rare cases, however, the course is rapid and death occurs in three or six months, or in one or two years. In the common form of the disease in which the duration is long, remissions may occur. Usually the improvement is not permanent and the disease ultimately advances to a fatal termination. Often the increase of symptoms follows over-strain, cold, or parturition. But in rare cases the remissions have been so long and so marked that a practical recovery has appeared to have occurred. Such cases of apparent recovery have been recorded by Charcot (one), Oppenheim (three), Byrom Bramwell (four) and Maas (one).

Treatment.—We have no means of curing the disease. Rest, tonic treatment, and good hygienic conditions are probably of slight service. I have tried the action of a large number of drugs, but have never been able to note beneficial effects from any, except perhaps from quinine. In several cases whilst the patient has been taking this drug distinct improvement has occurred : but it has not been possible to be quite sure whether this has been spontaneous or due to the action of the drug.

Bladder troubles and bed-sores require the same treatment as in myelitis.

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SECTION X

DISEASES IN WHICH ATAXIA IS A PROMINENT SYMPTOM

We have now to consider an important group of cases in which ataxia is a prominent symptom when the disease is fully developed. At the early period, however, or in a mild form of some of the affections placed in this group, ataxia is absent.

Spinal Diseases causing Ataxia.

Locomotor ataxia (tabes dorsalis).
Friedreich's disease (hereditary ataxia).
Ataxic paraplegia.
Combined postero-lateral degeneration associated with anæmia and various toxic conditions.
Some forms of spinal syphilis (Erb's spinal syphilitic paraplegia).
Occasionally disseminated sclerosis.

[Other diseases causing ataxia in which the primary lesion is not in the spinal cord: Tumours and other lesions of the cerebellum. Hereditary cerebellar ataxia (of Marie). The acute ataxia of Leyden (due to disseminated inflammatory patches in medulla, pons, and crura. Occasionally tumours of the prefrontal region. Internal ear disease. Rare forms of peripheral neuritis (ataxic forms). Occasionally hysteria.]

LOCOMOTOR ATAXIA: TABES DORSALIS

(Greek: *ἀ*, negative; *τάξις*, order. Latin: *tabes*, decay)

Etiology.—Tabes is the most common disease of the spinal cord. It occurs much more frequently in males than females—proportion 9 to 1 (2·7 to 1 in hospital practice, 25 to 1 in private practice—Mendel). It is a disease of the middle period of adult life, the onset being usually between thirty and forty-five. Before the age of twenty-five it is very rare, though cases of juvenile tabes are occasionally met with. The disease is more common amongst the inhabitants of towns than in rural populations.

The most important known factor in the causation of tabes is previous *syphilitic* infection. Statistics have shown (1) that a large proportion of the cases of tabes have been previously infected with syphilis; and (2) that in very few cases could previous syphilitic infection be excluded with certainty. Attention was first drawn by Fournier to the

relation between tabes and syphilis; Gowers and Erb soon afterwards published statistics in support of Fournier's view. The subject is still debated, though numerous statistics have been published in support of this relationship.

In favour of the view, that syphilis plays an important part in the causation of tabes, the following facts may be mentioned :—

A *history* of previous *syphilitic infection* is obtained in a large per centage of cases of tabes. This percentage has varied in different statistics, but the higher estimates have been 93 per cent., Fournier; 89·45 per cent., Erb; 80 per cent., Gowers; 90 per cent., Strümpell; 97 per cent., Dejerine.

Erb's latest statistics are based on 1,100 *cases of tabes in men* in the upper classes of society: a history of syphilis was obtained in 89·45 per cent.; no history of syphilis could be obtained in 10·54 per cent. 62·9 per cent. had presented definite secondary syphilitic symptoms, but 26·54 per cent. had had a chancre only. In many cases in which no history of syphilis could be obtained, there was a *possibility* of syphilitic infection; and in only 2·8 per cent. could the history be regarded as entirely negative, i.e., as furnishing no evidence or suspicion of the possibility of previous syphilitic infection.

Control statistics showed that amongst 10,000 men of the *upper classes*, over the age of twenty-five, only 21·5 per cent. had suffered from syphilis; whilst in 78·5 per cent. there was no syphilitic history. Hence in tabetic patients a history of syphilis is obtained nearly four and a half times more frequently than in non-tabetic men of the same classes of society.

The history is less reliable in men of the *lower classes* and in *women*. Thus Erb obtained the following results: In 158 male tabetic patients of the *lower classes* a history of syphilis was obtained in 77·2 per cent.; there was no history of syphilis in 22·8 per cent. In 1,300 non-tabetic men over twenty-five there was a history of syphilis in 6·54 per cent.; there was no history of syphilis in 93·46 per cent. Amongst 6,000 male patients suffering from diseases of the nervous system, Erb found that only 20 per cent. had suffered from syphilis.

Briefly stated, the results of Erb's statistics are that amongst 100 tabetic patients 80–90 have been infected with syphilis; whilst amongst 100 non-tabetic men only 20 per cent. have been infected with syphilis.

At a meeting of the London Pathological Society (1900) Sir William Gowers stated that amongst 100 consecutive male cases of tabes, in private practice, he had obtained unquestionable evidence of past syphilis in 68; in 12 there was a history of a chancre of uncertain nature, and syphilis was probable; so that in 80 of the cases syphilis was certain or probable. In the remaining 20 syphilis was *possible*, i.e. there had been exposure to the risk of infection, and in most cases several attacks of gonorrhœa. In none of the 100 cases of tabes could syphilis be excluded by the absence of any exposure to its risks. More recently

Gowers has stated that, excluding the medical profession, he has "never seen a case of true tabes in a man who has never run risks in the ordinary way."

Amongst 130 cases of tabes Collins found a definite history of syphilis in 70 per cent., and in 10 per cent. previous syphilitic infection was "possible." As contrast statistics Collins took "the histories of 140 cases of nervous diseases without tabes, seen in private practice, in which exactly the same inquiries and similar investigations were made to detect syphilitic infection." But he found only 8·3 per cent. had had syphilis. In other words, he found that tabetic patients have had syphilis at least eight or nine times more frequently than patients with other nervous diseases (excluding general paralysis of the insane).

As regards the absence of a history of syphilis in some cases of tabes, it is important to remember that occasionally such a history cannot be obtained in true syphilitic diseases. Pernet has recorded statistics respecting 124 cases of undoubted syphilitic skin diseases. The percentage of cases in which a history of syphilis was obtained was the same as in tabes. Hirschl has also found that in 63 cases of undoubted gummatous affections no history of syphilis could be obtained in 36·6 per cent.

Strümpell states that he has never seen any typical cases of true tabes in which previous syphilitic infection could be excluded with certainty.

Similar figures have been published by a number of other observers, but in some statistics the percentage of cases with a syphilitic history has been very much lower than those just mentioned. A comparison of the percentage of cases of previous syphilitic infection amongst tabetic patients with the percentage amongst cases of chronic spinal degenerative diseases (such as disseminated sclerosis) at the same hospital will usually demonstrate clearly, by the high figures in the former and the low figures in the latter, the relation of syphilis to tabes. Statistics show also that tabetic patients have suffered almost twice as often from gonorrhœa as non-tabetic patients of the same classes of society.

Tabes in Women.—Men suffer from syphilis much more frequently than women, and as already mentioned tabes occurs much more frequently in men. Syphilis is rarer in women of the upper classes than in women of the lower classes, also tabes is rarer in women of the upper than in women of the lower classes. Fehre has tabulated 305 cases of tabes in women (41 of his own cases, and the rest from published records). Definite evidence of syphilis was obtained in 154, and syphilis was very probable in 38. Considering the difficulty of obtaining evidence of syphilis in female patients, these figures strongly support the view that tabes is often indirectly the result of syphilis. Amongst 31 tabetic women recorded by Erb, 15 belonged to the higher classes, and 16 to the lower classes. Amongst those belonging to the higher classes, a definite history of syphilis was obtained in 60 per cent.;

a probable or almost certain history of syphilis in 33·3 per cent. Amongst those belonging to the lower classes a definite syphilitic history was obtained in 25 per cent., a probable syphilitic history in 31·2 per cent. and a possible history in 12·5 per cent. ; no evidence of syphilis could be obtained in 31 per cent.

Conjugal Tabes.—There are on record a number of cases of tabes affecting both husband and wife. In nearly all of these cases there has been a history of syphilitic infection. Hudovernig has collected 28 instances of married couples suffering from tabes : in 27 there was a distinct history of syphilis. Both man and wife have been affected with syphilis, or one has infected the other, and both have ultimately developed tabes.

Erb and others record cases in which several individuals, infected with syphilis from the same source, have developed, at a later period, tabes, or tabes with general paralysis, or cerebral or spinal syphilis.

These facts raise the question whether there is not a form of syphilis which is specially liable to lead to tabes, general paralysis, or syphilitic affections of the nervous system.

Juvenile Tabes—Rare cases of tabes are on record in which the disease has commenced in childhood, youth or early adult life. In these cases of *infantile or juvenile* tabes the history has usually shown that the parents of the children have suffered from syphilis, or the children have presented signs of hereditary syphilis ; in other cases the children have been infected with syphilis (extra-genital infection) at an early period of life—often through their nurses.

In 21 cases collected by Linser there was a clear history of syphilis in the parents in 17, in 2 syphilis was probable, in 2 doubtful. In 3 cases of juvenile tabes which I have recorded there was clear evidence of congenital syphilis in 2, in the third case the father had suffered from syphilis (*see* p. 315). In 46 cases collected by Hertz and Lemaire there was evidence of acquired syphilis (at a very early age) in 4, of hereditary syphilis in 26, and in 8 it was probable.

Oppenheim states that he has seen several cases of tabes in adults in which there was no history of syphilitic infection, but the father had suffered from syphilis or tabes.

Trevelyan records an instance of family tabes—the husband, wife and daughter all suffered from the disease.

In *support of the causation of tabes by syphilis* is the interesting fact that tabes is most common amongst the classes of people who most frequently suffer from syphilis, as, for example, military officers ; whilst tabes is very rare amongst clergymen and ministers of various churches, who suffer less frequently from syphilis than other classes of society. In Erb's table, giving the occupation in 550 cases of tabes, only one patient was a clergyman, and he had suffered from syphilis. Minor has pointed out that, in Russia, Jews suffer much less frequently from tabes than Russians, though Jews are very liable to suffer from other

nervous affections. He believes that the infrequency of tabes amongst Russian Jews is due to the fact that they rarely suffer from syphilis.

Whilst tabes only rarely occurs in persons who have not been infected with syphilis, it must be admitted that syphilis *alone* is usually not sufficient to cause tabes. Only 1 to 5 per cent. of persons infected with syphilis suffer from tabes. Other causes or factors probably aiding in the production of tabes are cold, injury, sexual excess, inherited or acquired weakness of the nervous system, many infectious diseases, excess in the use of alcohol and tobacco, and overwork and overstrain in various forms.

Erb believes that tabes is undoubtedly, in by far the majority of cases, a syphilogenic disease; but that it is not yet proved with certainty, though it is highly probable, that it is so in all cases; or in other words, we may almost say that only those persons who have been previously infected with syphilis are in danger of becoming tabetic later.

Many points in the etiology of tabes may be explained by the relation of the disease to previous syphilis. It explains the greater frequency of the tabes in males, the onset of the disease in the middle period of adult life, the frequency of the disease in certain classes of society, and its rarity in other classes. When the disease occurs in strictly moral married females syphilitic infection has usually occurred through the husband. When tabes occurs in very young or very old people, it is usually owing to syphilitic infection having occurred very early or very late in life, or in the former cases it is often due to congenital syphilis.

When tabes follows acquired syphilis, the onset in most cases is between six and fifteen years after the syphilitic infection, though it may commence as early as one year or as late as twenty years after the infection.

With reference to the relationship of tabes to syphilis there are, however, several points which require further consideration, because they have been put forward as *evidence against the syphilitic origin* of tabes.

The fact that only a small percentage, 1 to 5 per cent., of persons who are infected with syphilis develop tabes is no evidence against the relation between the two diseases, since, as Erb points out, only a small proportion of syphilitic patients develop gummata, and yet the syphilitic origin of the latter lesions is undisputed.

In some statistics the percentage of cases of tabes with a syphilitic history has been much lower than in the statistics of Erb and Gowers.

It is somewhat remarkable that in tabetic patients the symptoms of previous syphilis have been usually mild. Also tabetic patients do not often present signs of syphilitic disease of other organs. On post-mortem examination of tabetic patients definite evidence of tertiary syphilitic disease is not often met with. Still a number of cases are on record in which definite syphilitic lesions have been present in the brain, spinal cord or other organs, in addition to the tabetic sclerosis of the posterior columns. (In 15 out of 61 cases—Westenhoeffer.)

Lesser has published statistics as to the syphilitic lesions found post-mortem in cases of tabes. In 96 cases of tabes syphilitic lesions were found post-mortem in 27, i.e. in 28 per cent.; in all other autopsies, after the age of thirty-five years, syphilitic lesions were found in only 9·5 per cent.

In cases of syphilis in which there is severe ulceration, it is stated that usually tabes does not follow. In Herzegovina, Bosnia, Abyssinia and Central Asia, where syphilis causes such severe ulceration of the skin, it has been stated that tabes seldom occurs. But recent observations tend to show that in these countries tabes is more common than was formerly stated. It is also said that prostitutes seldom suffer from tabes.

Motschutkowski has recorded four cases of tabes in puellæ intactæ; also in seven of his cases the patient acquired syphilis after the onset of the tabetic symptoms. It is possible that some of these have been cases of syphilitic re-infection.

In spite of this supposed negative evidence, the facts stated at the commencement of this section show clearly the relation of tabes to syphilis in a large proportion of cases; but it must be allowed that in a small percentage of cases no syphilitic history can be obtained (about 10 per cent.).

Hitzig and Buzzard have suggested the possibility of a virus, capable of causing tabes, being associated with a soft chancre.

Accepting the view that there is a relationship between tabes and syphilis, there are three theories as to the exact connexion of the two affections—(1) that tabes is caused by the syphilitic poison; (2) that a toxin is produced in the system as a result of syphilis, and that this post-syphilitic toxin is the cause of tabes; (3) that syphilis is simply a predisposing cause of tabes, and that it produces an impairment of the nervous system and so renders it liable to degenerate from causes which would produce no effect on the healthy nervous system.

The changes in the spinal cord in tabes are not truly syphilitic in nature; they do not resemble those of tertiary syphilis; they must be regarded, not as syphilitic, but as post-syphilitic or para-syphilitic. Also anti-syphilitic treatment has not usually any curative action.

Strümpell has suggested that tabes is due to the action of a chemical poison or toxin, which develops in the system owing to the previous syphilitic infection. This poison he believes to exert its influence on certain sensory nerve fibres. The toxin of diphtheria acts in a similar manner, producing peripheral neuritis. The two processes are not quite analogous, since in tabes the change is usually progressive, more or less, for many years. Strümpell suggests that the toxin first produces a pathological change in the nervous system, and that complete atrophy and arrest of function is due to a wearing out of the nerve elements in the normal continued action.

In rare cases tabes has been attributed to injury (concussion of the

spine), exposure to wet and cold, over-strain, alcoholic and sexual excess. In many of these cases, probably, early symptoms of the disease have been already present, and these have been increased by the factors mentioned. In other cases, probably, these supposed causes have merely aided the action of some other cause.

Edinger has shown that in rats a degeneration of the posterior columns and of the posterior roots, similar to the changes in tabes, can be produced by prolonged and severe muscular exercise; and that these changes can be produced much quicker if the animal is rendered anæmic by the administration of pyridine.

According to Edinger, if the supply of nutrition to any nervous structure be defective, and if the work required from this structure be excessive, degeneration will occur. The previous syphilitic infection in tabetic patients is a predisposing cause of defective nutrition. Many tabetic patients are badly nourished, and often there is a history of over-work or sexual excess.

I have had interesting cases of tabes in which there has been great overstrain of the leg muscles through the working of a machine with the feet for 10½ hours daily, through walking long distances daily, etc.; in another case there was great overstrain of the arms in the work, and ataxia was especially well marked in the hands; in other cases there has been great overstrain of the eyes, and optic atrophy has developed. In all of these cases there has been a history or strong suspicion of syphilis.

Tuczek has shown that in chronic ergot poisoning symptoms and pathological changes similar to those of tabes can be produced, but the disease is not progressive.

Syphilitic Pseudo-tabes—Spinal syphilis occasionally produces symptoms which, at least in one stage of the disease, resemble in some points those of true tabes. But the complete or partial recovery, or the results of pathological examination show that the case is not one of genuine tabes.

Symptoms.

The symptoms of tabes dorsalis are numerous and varied, but certain of them are present in the majority of cases, and a summary of the more important symptoms in the three stages of the disease will be of service.

1. In the early or *pre-ataxic* stage the most important symptoms are shooting pains in the legs, loss of the knee-jerks, and loss of the reflex action of the pupils to light, whilst reaction to accommodation is preserved (Argyll-Robertson pupil). When these three symptoms are all present the diagnosis is certain. Along with loss of the knee-jerks there is usually loss of the tendo Achillis reflex, and the Achillis jerk is often lost before the knee-jerk. Other symptoms in this stage are—paræsthesia, numbness of the soles of the feet, numbness and tingling in the

little and ring fingers, a girdle sensation, bands of diminished tactile sensation on the trunk, muscular hypotonus, temporary paresis of ocular muscles causing diplopia and squint. Sometimes slight bladder symptoms, impotence, gastric crises, joint affections, and analgesia of the legs occur in this stage. In one variety of the disease optic atrophy, producing progressive failure of vision, is an early symptom. Sometimes there is loss of the vibrating sensation (" bone sensibility ") at an early period of the disease. This stage may last for many years before ataxia occurs. In rare cases the ataxia appears early.

2. In the second or *ataxic* stage, muscular inco-ordination develops in the legs. There is inability to stand with the feet together and eyes closed (Romberg's symptom). Walking becomes more and more difficult, and finally impossible. The movements of the arms are also often ataxic. Marked sensory disturbances of various kinds develop, and joint affections and trophic changes may occur.

3. In the last or *paralytic* stage the patient is no longer able to walk, he is confined to bed, and weakness of the legs may finally develop; but true paraplegia is very rare. Bed-sores, cystitis and pyelitis often occur at the termination of the disease.

After this sketch of the clinical course the symptoms may be considered in separate groups.

Ataxia or inco-ordination of movements is one of the most characteristic symptoms of tabes, though other symptoms are usually present, for many years before it is detected. In very rare cases, however, ataxia has been observed as an early symptom of the disease. The ataxia of tabes is made worse when the eyes are closed. The muscular power of the affected limbs is unimpaired, or it is affected only late in the disease or when some complication is present.

Usually the ataxia is first noted in the legs. A commencing ataxia can be detected by asking the patient, when seated, to touch the knee of one leg with the heel of the other. The moving leg is carried to one side or beyond the point aimed at, and several attempts may be necessary before the heel touches the knee. The patient being seated on a chair, the inco-ordination is also noticed when he attempts to cross his legs, or to describe a circle in the air with his great toe, or to touch an object with it.

The early unsteadiness is noticed when the patient attempts to walk backwards; when he is suddenly told to turn round or to halt whilst walking; when he is going downstairs; when he suddenly stands up and walks; and when he attempts to stand on one leg or to hop. Rosenbach has pointed out that, even at an early stage, there is often inability to stand on the tiptoes, especially if the eyes are closed.

When the patient brings his feet together (heels and toes being in contact) there may be inability to stand if the eyes are closed: he sways

from side to side, or backwards and forwards, and would fall over if not supported (Romberg's sign). This unsteadiness is well brought out in slight cases by asking the patient to stoop down, and then assume the erect posture with the eyes closed (Oppenheim).

At first the patient is able to stand with his feet together if the eyes are open, but not when the eyes are closed. Later even with the eyes open, bringing the feet together causes him to sway about, and he has to separate his feet a little to prevent himself from falling. Often the unsteadiness is first noticed when the patient is walking or standing in the dark, or when he closes his eyes whilst washing his face in the morning.

With regard to the aid of vision in arresting or diminishing the un-

(a)

(b)

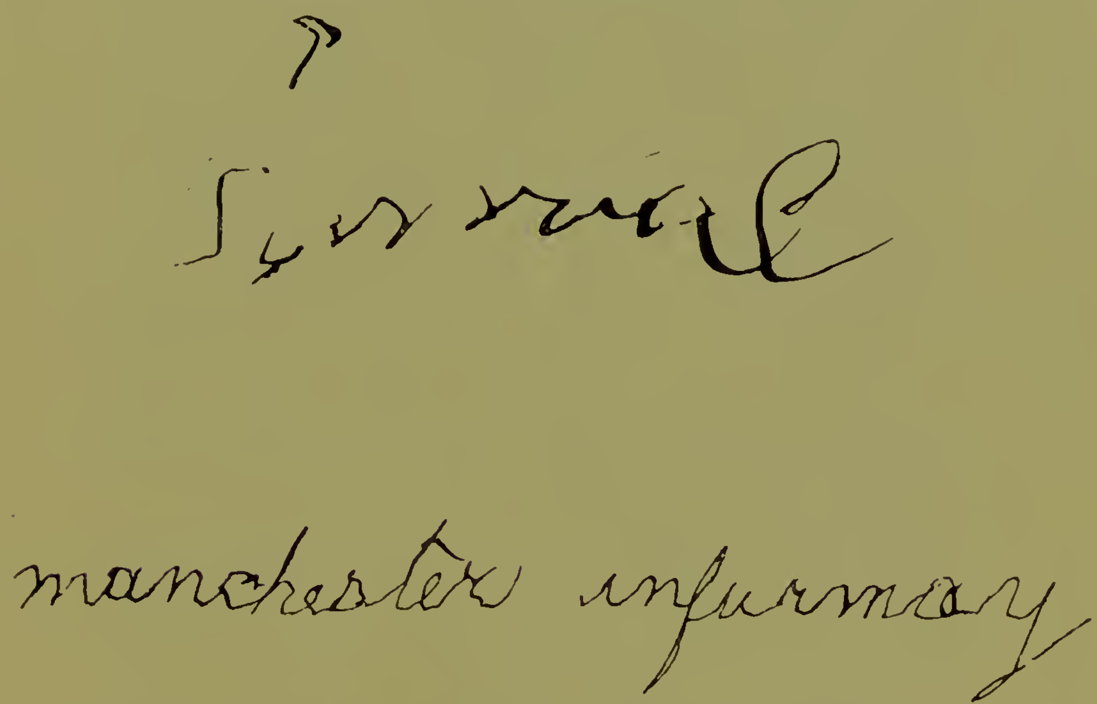

FIG. 132.—Handwriting in Tabes Dorsalis. (*a*) Attempt to write name "Samuel." Case with marked ataxia of arms. (*b*) Writing of a patient suffering from slight ataxia of the arms.

steadiness, it has been pointed out that it is not necessary for the patient to see the feet. Opening the eyes checks the tendency to sway and fall when the room is very feebly lighted. Also when a patient is quite blind from optic atrophy, Romberg's sign may be observed, the patient becoming unsteady on closing the eyes, although with the eyes open he can stand steadily.

When standing, the knees are sometimes a little over-extended (bent a little backwards). In time the gait becomes unsteady. In walking the feet are raised too high and thrown too far forward, the heels are often brought down very suddenly with a stamp, and the steps are of unequal length. Often there are irregular deviations of the legs laterally, and the patient walks with his feet apart and looks down at them.

Occasionally there is a sudden weakness or giving way of the legs, which causes the patient to stumble or fall. The gait in course of time becomes more and more uncertain and irregular; the patient is unable to get about without the aid of a stick; later he must be supported on one or both sides by an attendant; finally he is unable to stand without assistance; and in the last stage of the disease the ataxia becomes so great that he is confined to bed.

In the arms the ataxia is usually less and appears at a much later period, but in a few cases (cervical tabes) the arms are chiefly affected. The inco-ordination, unsteadiness, or clumsiness is noted in performing fine movements, such as buttoning the clothes, writing, playing the piano, touching any object (as the tip of the nose) with the index finger, bringing the tips of the two index fingers suddenly together after the hands have been widely separated, picking up a pin or small object from the table. This inco-ordination, or clumsiness is noticed also when the patient is using a knife and fork or spoon, or drinking from a glass; finally as it advances he may be unable to feed himself. The inco-ordination in all of these movements is more marked (at first only seen) when the eyes are closed.

Occasionally involuntary movements of the fingers are noticed when the hand is at rest and supported—sudden rising and sinking (extension and flexion) of the fingers. At first this only occurs when the eyes are closed.

Occasionally ataxia in the movements of the tongue, face and vocal cords has been observed.

Hypotonus.—Often there is a remarkable flaccidity of the limbs, or loss of muscular tone on passive movements (though there is no loss of muscular power). When the patient is in the horizontal position the examiner is able to move the limbs with less muscular resistance than is felt in the normal condition; also the movement can be carried out to a much greater extent than in health. For example, in passive movement the leg can sometimes be flexed at the hip to an abnormal degree, so that the foot approaches the head, or even almost touches it. This is due to diminished muscular tone (or hypotonus).

Frenkel has drawn attention to the diminished muscular tone (hypotonus) which is noted on active movements in tabes, though the muscles appear normal and present no signs of paresis. The tabetic patient can often perform movements to an abnormal extent at the various joints. When a healthy man is placed in the horizontal position (on a couch) he cannot raise the leg very high, if the knee be kept extended. Usually he cannot raise it to an angle of more than 65° with the horizontal line (couch level).[1] If raised higher, the knee must be flexed. This is owing to the resistance of the semi-membranosus, semi-tendinosus

[1] In fifty non-tabetic individuals (either healthy persons or patients not suffering from diseases of the nervous system) I found that forty-four could not raise the leg to an angle of more than 65° with the horizontal line.

and biceps at the back of the thigh. In tabes, however, the leg (fully extended at the knee) can often be raised to an angle of 80° or 100° or more from the couch. This is owing to loss of tone (hypotonus) of the muscles just mentioned. The angle to which the leg can be raised may be measured by a graduated semicircle and a long piece of string as described on p. 13. On account of the hypotonus of the adductors, the thighs can be abducted (when the patient is on the couch) until both the legs are almost in one line with the couch. Owing to

Fig. 133.—Tabes Dorsalis, showing abnormal position to which the leg can be raised (about 120° from the horizontal level) owing to muscular hypotonus.

muscular hypotonus the knee can often be flexed until the heel touches the tuberosity of the ischium; the knee can be over-extended; the spine can be flexed when the patient is on the couch until the head comes in contact with the couch between the legs; and other abnormal movements can be performed.

In the early and pre-ataxic stage muscular hypotonus is often present. I have found it of much value in diagnosis as a confirmatory sign in early tabes (when other symptoms have been slight and few), and especially in the form of tabes commencing with optic atrophy.

As already mentioned, the ataxia is not usually associated with any

muscular weakness, and even when the inco-ordination has become extreme, the movements of the legs and arms are quite powerful. Electrical examination in uncomplicated tabes shows no reaction of degeneration and no other electrical changes of importance in the muscles and nerves.

The Reflexes.—The loss of the knee-jerk in tabes was first pointed out by Westphal, and hence this symptom is spoken of as Westphal's sign. The knee-jerk is usually lost on both sides; this is one of the earliest and most constant signs of tabes, and may precede the onset of ataxia for many years. (When in doubt whether the knee-jerk is present or not, Jendrássik's method of testing the reflex should be employed. *See* p. 63.)

In any cases of suspected tabes, if the knee-jerk is not lost the greatest care should be taken before making a diagnosis of that disease, since in tabes the knee-jerks are almost always lost. In 271 cases of tabes Bonar found the knee-jerks lost in 258 cases, i.e. 95 per cent. Very rare cases are met with, in which for a time at least, at the earliest period of the disease the knee-jerks are present, though other symptoms are decided. Also in very exceptional cases, one knee-jerk is absent and one present just at first.

Though the knee-jerks are lost in tabes, direct muscular mechanical irritability, as shown by tapping the muscles of the legs, especially the quadriceps muscle, is always present.

Hughlings-Jackson and J. Taylor have pointed out that in tabes with loss of knee-jerks if an attack of hemiplegia should occur, the knee-jerk returns on the paralysed side. In very rare cases the knee-jerk has returned temporarily when there has been no hemiplegic attack (Berger, Donath).

Babinski has pointed out that the tendo Achillis reflex is usually lost along with the knee-jerk in tabes. But the former reflex is usually lost before the latter, and in the rare cases of early tabes when the knee-jerks are at first present, the tendo Achillis reflex is lost, and this sign may then aid in the diagnosis.[1] Occasionally the tendo Achillis jerks are present when the knee-jerks are lost.

The cutaneous reflexes—plantar, cremasteric, abdominal and epigastric—are often increased at the early stage. In the later stages of the disease, or when tactile anæsthesia occurs, the cutaneous reflexes may be lost. (Rosenbach has drawn attention to the increase of the abdominal reflexes in association with loss of knee-jerk in tabes. I have often observed the same condition in diabetes mellitus.) The plantar reflexes are of the normal flexor type.

[1] Thus in a case of tabes which I reported (*Medical Chronicle*, August, 1902), the symptoms were—double optic atrophy, ataxia, Romberg's sign, Argyll-Robertson pupil, muscular hypotonus, girdle pain, shooting pains in the legs, numbness in the feet, zone of diminished tactile sensation on the right side of the chest. The *tendo Achillis reflexes were absent, but for sixteen months the knee-jerks were present*: at the end of that time they disappeared.

Paralysis and atrophy of muscles of the limbs are occasionally met with in tabes. But these are to be regarded as very rare and unusual complications; and tabes, apart from these complications, is a disease which does not cause muscular paralysis in the limbs, except at the terminal stage of the disease. Even at the last stage loss of power is often incomplete and would be better described as paresis, also it is frequently temporary.

Hemiplegia occasionally occurs as a complication owing to cerebral lesions—hæmorrhage, thrombosis or embolism. Previous syphilitic infection has probably, directly or indirectly, played some part in the causation of many of the first two affections. A syphilitic gumma may also produce hemiplegia or monoplegia. A very rare complication has been paraplegia due to acute or sub-acute myelitis (probably of syphilitic origin).

But in addition to these forms cases of localised paralysis of muscles of the limbs have been recorded, such as paralysis of the muscles supplied by the peroneal, musculo-spiral and median nerves, paralysis of the adductors of the thigh, and paresis and paralysis of the legs. Some of these affections have probably been cases of pressure paralysis from accidental compression of nerves, in other cases the paralysis has been due to motor neuritis.

In very rare cases muscular atrophy has developed. Usually this has occurred at a late period of the disease, chiefly in the legs and arms. The muscles supplied by the peroneal nerves have been most frequently affected. The atrophy slowly increases, but is said not to have the tendency to spread slowly from one group of muscles to another, as in progressive muscular atrophy. Fibrillary contractions are sometimes present, sometimes absent.

Talipes equino-varus, with marked flexion of the toes, has been recorded in a considerable number of cases (tabid club-foot of Joffroy). This condition has been attributed to peripheral neuritis, but probably the pressure of the bed-clothes on the dorsum of the foot has assisted in the production of the deformity.

Sensory Symptoms.—*Pain.*—Most tabetic patients suffer from pains; though they vary in intensity, they are seldom entirely absent. They are often the first and most prominent symptoms, and may precede other symptoms for many years.

Following the description of Sir William Gowers, the pains of tabes may be divided into—(*a*) pains which are commonly momentary, but which succeed each other after a short interval in the same place, frequently for hours, sometimes for days; (*b*) pains which are prolonged.

As regards the first form, (*a*), the patient usually suffers from paroxysms of pains, but each pain is of very short duration; it is sudden or darting in character like an electric shock (lightning pains). The pains are repeated frequently for a few minutes or for some hours, or they may be repeated for a day or two. Then the patient is free from

pain for days, weeks, or months. The pains occur chiefly in the legs and lumbar region. In the arms they are much less frequent and less severe, except in cases of cervical tabes. Pain may radiate round the trunk, and produce a girdle sensation or feeling of a band or constriction round the waist. This is often an early symptom of the disease. Occasionally the bladder and rectum are the seat of the pains.

The momentary pains are of two kinds—(1) deep pains which appear to be in the muscles or bones, and are compared to a feeling as if a knife were being thrust into the flesh and turned round; (2) superficial pains of lightning character, which are referred to the skin. In time the skin may become very tender, so that the patient cannot bear the contact of the bed-clothes or other objects at the affected part; sometimes the patient is hypersensitive to cold and heat in the region of the pain.

(*b*) The pains which are prolonged last for hours, days or longer in the same place; they present variations in intensity, but not the complete intermissions or considerable remission of the first class. Often the pain is very severe and burning or dragging in character. In other cases the prolonged fixed pains are widely diffused, but not severe; they continue for weeks or even months without ceasing; they often cause sensations of numbness, tingling, swelling, heat, or cold.

It is only the sharp momentary pains of the first class (*a*) which are characteristic of tabes.

In some cases the pains of tabes are of moderate severity, the paroxysms seldom, and duration of the attacks short; in other cases the pains are very violent or frequent.

The pains of tabes are increased by cold and damp. Patients suffer more from the pains in a cold climate than in a warm one; more in winter than in summer; and more in a damp climate than in a dry one.

The pains of tabes differ in their localisation from those of neuralgia. Tabetic pains are in the region of skin supplied by a segment of the spinal cord; true neuralgic pain is in the cutaneous distribution of a definite peripheral nerve.

The tabetic patient often complains of tingling or " pins and needles " in the limbs, especially in the soles of the feet. Also the feet may feel as if covered with cotton wool, or there may be a sensation of cold or heat in the legs.

Numbness and tingling may be met with in the region of distribution of the ulnar nerve, or of the eighth cervical and first dorsal nerve roots (*see* Fig. 134). Biernacki has pointed out that in tabes (and also in general paralysis of the insane) often strong pressure on the ulnar nerve, just behind the inner condyle of the humerus, does not cause pain; whilst in health, pressure on the nerve at this point gives rise to the well-known painful sensation. In half of his cases there was also analgesia of the skin in the region of the ulnar distribution. Sarbō

has pointed out that the peroneal nerve, just behind the head of the fibula, is also frequently analgesic on strong pressure.

Objective Sensory Symptoms.—In the majority of cases of tabes some objective affection of cutaneous sensibility is found. But all forms of sensation are rarely affected equally; often one or two forms only are affected and other forms are spared: at the early and pre-ataxic stage no affection of cutaneous sensation may be detected.

The observations of Hitzig, Laehr, Frenkel and others have shown that even in the early stages of tabes there is often a peculiar localised diminution of sensation to light tactile impressions on the trunk, but there is very rarely total anæsthesia. Often the sensations for temperature and pain are intact, or only affected in patches, in the region of the diminished tactile sensation. Hence in testing for these areas of diminished sensation the touch should be as light as possible, corresponding parts should be compared, and the object with which the examination is made should have the same temperature as the patient's skin. Frenkel recommends the examination to be made with the tip of the finger warmed in hot water, if necessary, so as to have the same temperature as the patient's body.

The areas of diminished sensation on the trunk usually take the

Fig. 134.—Zones of Diminished Cutaneous Tactile Sensation. Tabes dorsalis. In the central figure there was also a strip of diminished sensation down the inner side of each arm and hand.

form of bands or girdles around the chest or abdomen. They are often bilateral, in other cases unilateral. The region supplied by the mid-dorsal nerves is most frequently affected, especially the region of the fourth intercostal space and lower part of the scapula. The vertical extent of the bands varies considerably. Very often the patient is ignorant of the diminution of sensation until the examination is made. The bands or zones of diminished tactile sensation do not correspond to the distribution of single nerves, but to the distribution of spinal nerve roots. In advanced cases the diminished sensation extends down

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the inner (ulnar) side of the upper arm and forearm and into the little finger. Finally the whole arm may be affected. At the border of the zone of diminished tactile sensation, and in the region between adjacent zones, there is frequently hyperæsthesia for cold.

Frenkel found that sensation to pain was also affected on the trunk in a few cases, but this symptom developed after the partial tactile anæsthesia.

The zones of diminished tactile sensation are present also in other diseases, such as syringomyelia; nevertheless they are of diagnostic service in early tabes and in cervical tabes, when other symptoms are few. They are of value in the diagnosis between true tabes and pseudo-tabes (ataxic form of multiple neuritis) since they do not occur in the latter affection.

Marinesco has pointed out that there are four chief foci of anæsthesia in tabes : (1) thoracic—zones of diminished tactile sensation in the region of the nipple, as just described ; (2) in the external genital organs ; (3) in the legs, especially the feet ; (4) down the inner side of the arm, forearm and hand. These four regions are the parts in which subjective symptoms first occur—i.e. pains or numbness, etc.

Longer known than the diminution of tactile sensation, is diminished sensation to pain ; it is best marked in the legs, and is often an early sign. According to Frenkel and Foerster, in the legs the sensation for pain is affected before tactile sensation, whilst in the arms and trunk the reverse condition occurs. A sharp pin prick is not distinguished from the head of the pin or the touch of the finger in those cases in which the pain sensation is lost. When the pin prick is of longer duration it is sometimes felt first as a tactile sensation, and only after a few seconds is the painful sensation felt (double sensation). In advanced cases a pin prick of long duration, or the stimulation of the faradic battery, may cause no pain in certain parts of the skin.

In addition to the zones of diminished tactile sensation on the trunk, there is often diminution or loss of the sensation to touch or pain in the feet, and occasionally in the hands. Anæsthesia and analgesia are occasionally localised to the distribution of peripheral nerve trunks.

Occasionally one pin prick gives rise to five or six or more sensations of pain. Occasionally no sensation of pain is produced by strong faradisation of muscles.

The conduction of pain is frequently delayed ; this is a common early symptom. Tactile and temperature sensations are only occasionally delayed. A pin prick may be felt only after an interval of 2 to 5 seconds. Sir William Gowers records a delay of 7 seconds, and Eulenberg 15 seconds. There may be also an " after pain " lasting for many seconds, or the pain may not reach its greatest intensity for several seconds after it is first felt ; occasionally there is a second or third maximum.

The power of localising a tactile sensation may be impaired, lost or

perverted. A touch at one point is referred to a part a short distance away. A touch or prick on one leg may be referred to the other, and often to a corresponding part (allóchciria). A prick on one part of the leg may be felt in several places (polyæsthesia).

The sense of temperature is not often affected alone, but it may be impaired when the sensation of pain is affected. The loss of temperature sensation may be complete (for both heat and cold) or partial (for heat or cold alone). According to Starr, the sensation for heat is often lost, whilst there is hyperæsthesia for cold. Occasionally there is loss of sensation to temperature and pain, whilst tactile sensation is spared or but little affected (as in syringomyelia). Often the painful sensation produced in a normal individual by compressing the testicles is absent. A peculiar form of anæsthesia is the loss of the sensation of tiredness after long exertion, which has been described by Frenkel.

Hyperæsthesia and especially hyperalgesia are sometimes met with, especially in patches. In these regions the slightest touch may give rise to a painful or burning sensation.

In exceptional cases marked anæsthesia to many forms of sensibility occurs—such as total anæsthesia of the limbs and trunk to tactile and painful sensations, with loss of muscular sense. Occasionally loss of the stereognostic sense occurs, and may be detected in the hands. The vibrating sensation is often lost on the feet or legs (*see* p. 80); this may be a very early symptom in the pre-ataxic stage. The vibrating sensation may be lost on the legs when no impairment of other forms of sensation and no ataxia can be detected.

An affection of the *sensation of passive movement* (joint sensation) can often be detected. (The passive movements should be exceedingly slowly made, the hand of the examiner must remain in the same position on the patient's limb during the movement, the pressure on one side must not be greater than on the other, and the limb should not come in contact with the bed-clothes; also the muscles of the limb should be relaxed during the movement.) A common affection is the inability to tell when the great toe is passively moved in the direction of dorsal or plantar flexion, whilst in health a slight degree of flexion or extension is at once recognised: or the patient may be unable to tell the direction of the movement. In some cases the sense of passive movement is not entirely lost, but the patient is only able to tell the nature of the movement when its extent is much greater than in health.

A symptom met with in severe cases of tabes is loss of the sense of position. The patient cannot tell the position of the limbs when the eyes are closed. This is owing to the diminution or loss of sensation in muscles and tendons. Also in some cases the patient is unable to distinguish between light and heavy weights.

Loss of the sensation of pain on severe pressure of muscles (especially of the calf muscles) is a common symptom (*see* p. 82). There is often also analgesia of the tendo Achillis.

Ocular Symptoms.—A very important symptom of tabes is the Argyll-Robertson pupil, i.e. when light is allowed to fall on to the retina suddenly there is no reflex contraction of the pupil, but the pupils contract on accommodation. In testing for this symptom the eyes should be directed to a distant object. One eye is shaded from the light by the hand of the examiner, and then, by suddenly removing the hand, light is allowed to fall on to the retina. The electric light is useful in cases where the reaction is doubtful or feeble. Another method of examination is by throwing light into the eye by means of an ophthalmoscopic mirror or lens. In health both pupils contract; in most cases of tabes the pupils do not contract when examined in this way. Usually the pupils contract to accommodation (when the eyes are directed to a near object). The Argyll-Robertson pupil is a very early sign of tabes, and may precede the ataxia for a long period. It is present in about two-thirds of the cases. It is usually present in both eyes, but occasionally the sign is unilateral, whilst in the other eye the pupillary light reflex is simply diminished. In some early cases the pupillary reflex is variable; at one period the pupils do not react to light, whilst at other times the reflex is present, but sluggish.

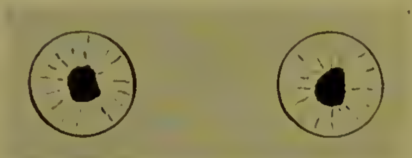

Fig. 135. — Pupils in Tabes Dorsalis: not circular (eccentric).

It is important to remember that though the pupils in tabes usually do not react to light, this is not always the case; and the fact that the pupils react normally to light does not exclude tabes in the differential diagnosis.

Sometimes the pupils are fixed and do not react either to accommodation or light, but they are not markedly dilated, as in paralysis of the third nerve.

In health when the skin at the back of the neck is painfully stimulated, as by pinching or by a faradic current, the pupils dilate. This reaction is often absent in tabes, but occasionally it is absent in health.

The pupils in tabes are often very small—myosis; sometimes they are of medium size; occasionally they are greatly dilated; sometimes they are unequal. Often they are irregular or not quite circular in outline or eccentrically situated, and these irregularities in the outline of the pupil often precede the Argyll-Robertson pupil.

Temporary or permanent paralysis of one or more ocular muscles may occur. The temporary paralysis occurs chiefly in the early stage, permanent paralysis in the advanced period of the disease. Strabismus and diplopia are produced by the paralysis of ocular muscles. The external rectus is most frequently affected, but the third nerve or one of its branches may also suffer, and unilateral or bilateral ptosis may be produced. In rare cases most or all of the ocular muscles become paralysed, and the condition known as ophthalmoplegia is produced.

The paralysis of ocular muscles is usually due to changes in the nerve fibres; in some cases it is due to nuclear changes.

Primary optic atrophy occurs in many cases; according to Strümpell in 10–15 per cent., according to Sir William Gowers 6·5 per cent. Both eyes are almost always affected, though one eye may be affected earlier and more markedly than the other. The optic atrophy is primary, i.e. there are no evidences of previous optic neuritis. The margins of the optic discs are sharply defined, and the colour of the discs gradually becomes paler, until at last they are grey or greyish white. The temporal half of each disc is said to be first affected. The optic atrophy usually progresses slowly, sometimes rather rapidly, and causes increasing failure of vision until total blindness is produced. But vision may not be entirely lost for ten or fifteen years; in other cases it is lost within six months. According to Marie, three years is the mean time in which vision becomes lost. There is often a period during the progress of the atrophy when the visual failure ceases to advance for a time. In the development of the visual failure, at first the periphery of the field suffers and colour vision, especially for green and red, is affected. There is concentric diminution of the field of vision: often the restriction of the field is irregular; sometimes there are sector-form restrictions; sometimes the temporal half is more restricted than the nasal. In some cases the acuity of central vision is preserved for a long period, and the visual defect consists simply in restriction of the field. In other cases acuity of central visions fails, with or without restriction of the fields. Central scotomata are quite exceptional, and should always be regarded as evidence against tabetic atrophy. (In disseminated sclerosis central scotomata are common.) Optic atrophy may be a very early symptom (2 per cent. of cases). Often there are only very few other symptoms of the disease, such as absent knee-jerks and shooting pains, when the patient comes under observation. Occasionally the only other symptoms for a long period are loss of the knee-jerks and tendo Achillis reflexes, and in rare cases the knee-jerks are present at first. Ataxia is usually very late in developing, and may not be detected for many years; in a large number of these cases it does not occur. In some cases optic atrophy appears after other signs of tabes have become well marked. In such cases the spinal symptoms often become stationary when optic atrophy develops; even diminution of ataxia and other symptoms has occurred after the optic atrophy has developed. To these general statements there are exceptions.

Mendel estimates that amaurosis is more common amongst tabetic women than men. Optic neuritis is not a true symptom of tabes. A few rare cases are on record, in which this sign has been met with in a tabetic patient; but probably it has then been due to some syphilitic brain affection complicating the tabes.

Affections of other Cranial Nerves and Special Senses.—The sense of smell is lost in rare cases. Occasionally deafness is observed, which

has the characters of nerve deafness. Occasionally there are attacks of vertigo with vomiting and noises in the ears—symptoms simulating those of Menière's disease.

The fifth nerve is sometimes affected, and in the region of the distribution of this nerve there may be pains, numbness and diminution or loss of the sensation for touch or pain ; sometimes the patient feels as if the face were covered with a mask or with parchment. Other rare symptoms are analgesia of the mucous membrane of the mouth, anæsthesia of the cornea, and falling out of the teeth (trophic changes). Rare trophic changes are neuro-paralytic ophthalmia, ulcerations in the mouth, and herpes zoster. In very rare cases the motor branch of the fifth nerve has been affected, and atrophy and paralysis of the muscles of mastication have been noted.

Paralysis of the muscles of the vocal cords is sometimes observed. The abductor muscles (posterior crico-arytænoids) are most frequently paralysed, and stridor on inspiration, or laryngeal crises may result. Sometimes the adductors are also paralysed. Occasionally all the muscles of one vocal cord are completely paralysed. Often the paralysis of laryngeal muscles can only be detected by laryngoscopic examination. (Pathologically degenerative changes have been found in the peripheral nerves and also in the medulla.)

In rare cases there is atrophy and paralysis of one-half of the tongue. Also on the same side frequently one vocal cord and one-half of the soft palate are paralysed. The condition remains unilateral, and never progresses to true bulbar paralysis. (In these cases atrophy and disappearance of nerve cells in the nuclei of the hypoglossal and spinal accessory nerves have been noted. Degeneration has also been present in the peripheral nerves supplying the parts mentioned.)

Bladder and rectal symptoms. In the early stages slight bladder symptoms are not uncommon. Micturition is often sluggish, and the patient has to wait for a minute or two, and has to strain before he can pass water ; in other cases there is occasionally dribbling of a few drops of urine. Not infrequently the bladder is not completely emptied, but absolute retention and absolute incontinence are very rare ; they are usually temporary, and usually occur only at an advanced stage of the disease. In the last stage of the disease complete retention of urine may occur, and may be followed by overflow incontinence. Cystitis may then develop, and may lead to pyelonephritis.

Constipation is common, owing to loss of power of the muscular coat of the intestines and rectum. In the last stage the sphincter ani may become weak, and incontinence of fæces may occur, but this is very rare.

The sexual functions are often affected. Sometimes impotence is an early symptom ; in other cases there is early sexual excitement. In the last stage frequently sexual desire is lost, and erection and ejaculation fail. Mendel estimates that sterility is three times more common

amongst women suffering from tabes than amongst non-tabetic women in the same social position.

Functional visceral disturbances, so-called "*crises*," often occur. The most common are gastric crises. In these attacks the patient suddenly suffers from obstinate vomiting, associated with severe epigastric pain or girdle pain; nausea is sometimes present, sometimes absent. The vomited substance at first contains food, afterwards it consists of clear fluid, and finally it contains bile; occasionally a little blood may be vomited. Often the vomited matter contains much hydrochloric acid, but not always. The pain and vomiting continue, though no food be taken. During the attacks the pulse is often rapid; the patient complains of palpitation and vertigo, and he is unable to take food. The tongue is usually normal. The attack continues for some hours or days, or for a week; the symptoms subside suddenly, sometimes gradually, and the gastric condition becomes normal. These crises occur at irregular intervals, sometimes frequently, sometimes seldom. Attacks may occur in which the pain is present but vomiting absent, or vomiting may occur without pain. (I have known gastric crises to be the first symptom of the disease to attract attention, and for some time this symptom and loss of the knee-jerks were the only evidences of tabes which could be detected.)

Fig. 136. Perforating ulcer of the foot: tabes dorsalis.

Occasionally there are rectal crises in which the patient suffers from paroxysms of pain in the rectum, with severe tenesmus, and a sensation of a foreign body in the rectum. Intestinal crises—attacks of diarrhœa with or without pain—have also been described. Sometimes constipation is a troublesome symptom.

Laryngeal crises are less frequent than gastric crises. In these attacks there is a sudden difficulty of breathing with stridor (inspiratory and expiratory). In other cases a paroxysm of coughing is the prominent symptom, and is associated with shortness of breath and cyanosis.

Rare symptoms are—renal crises, in which there are paroxysms of pain like renal colic; vesical crises, in which there are attacks of pain at the neck of the bladder; and urethral crises, in which the pain is in the urethra. In the female, crises have been described in which there is great sexual excitement—"clitoris crises."

Trophic and vaso-motor symptoms are occasionally present. Of these, the most important are perforating ulcers. They usually commence in a corn, which ulcerates at the centre. The ulcers are circular and deep, the margins sharply defined, and a slight purulent discharge is often present. The ulcers are situated chiefly on the balls of the toes,

on the soles of the feet, or on the heels. The severe forms may extend deeply to bones and joints, and may necessitate amputation. Often there is an area of anæsthesia round the ulcer. In the severe forms, healing is very difficult to obtain. Other trophic changes are thickening or falling off of the nails, falling out of the teeth, falling off of the hair, alteration in the growth of the hair, ecchymoses in the skin, atrophy of the skin, hyperidrosis and anidrosis, local sweating of the palms of the hands, of the soles of the feet, and of one side of the head.

Herpes zoster, small subcutaneous hæmorrhages, and gangrene are met with in very rare cases. Rupture of tendons and disappearance of intra-articular ligaments sometimes occur.

The bones may become brittle, and break easily through some slight strain or injury (so-called spontaneous fractures). In the union of these fractures a large quantity of callus is formed.

The X ray photographs often show that the bone shadow is not so deep as in the normal condition. This has been attributed to a rarefaction of bone tissue.

"*Charcot's joint disease.*" This is a somewhat rare complication. It may occur in the ataxic stage or as one of the earliest symptoms in the pre-ataxic stage. The onset of this affection is sudden ; the joint becomes rapidly swollen through distension with fluid. There is no pain and no elevation of temperature. Soon the adjacent parts, or the whole limb, may become swollen ; but there is no pitting, no sign of inflammation, and the skin is pale. After a few days the general swelling subsides, but the joint remains distended with fluid. Sometimes the joint returns to its normal size after a few weeks or months, but there is crepitation or crackling on movement, and relapses are apt to occur ; in other cases the joint remains swollen for a very long period. As the effusion is absorbed the cartilages become eroded, the bony surfaces of the joint become worn away, and osteophytes may form ; also the ligaments become relaxed. Owing to these two changes abnormal movements can be made laterally, or backwards, or in the direction of over-extension. Suppuration of the affected joint is quite exceptional. Bony excrescences and periarticular new bony formation may appear. The joints most often affected are the knee, hip, elbow, and shoulder, but other joints may suffer (Weizsäcker's statistics on 109 cases). Muscles around the affected joints often atrophy.

The tabetic foot is a condition due to disease of the tarsus. The onset of the affection is sudden ; within twenty-four hours a prominence develops on the dorsum of the foot, near the tarso-metatarsal joint ; it does not pit on pressure. The sole of the foot and arch of the foot become flattened, and the inner border of the foot swollen. There is no pain, but the mobility of the foot is impaired. Pathologically the changes consist in wasting, atrophy and disintegration of the bones of the tarsus and metatarsus; but there is no suppuration.

Apart from the joint disease the ligaments are often relaxed, and

hence the limb can be placed in abnormal postures. Occasionally a peculiar twisted position of the toes is seen.

In rare cases ossification of fibrous tissue connected with muscles has been observed. At a late stage bed-sores may form over the sacrum.

Usually the general nutrition of the tabetic patient is impaired; loss of weight is common, and sometimes there is anæmia. If by tonic treatment—good food, drugs, change of climate etc.—this failure of nutrition can be arrested, the symptoms of the disease will often diminish. Hence attention to the general health is important, and here we have a hopeful indication for treatment.

In rare cases the general wasting has been such a prominent symp-

FIG. 137.—Charcot's Disease of Knee Joint. Tabes dorsalis. Swollen joint diseased; smaller knee joint normal.

tom that the tabetic symptoms have been overlooked, and the case treated as one of wasting of unknown cause.

The *cerebro-spinal fluid* obtained by lumbar puncture usually contains a great excess of cells, which are lymphocytes. A large number of observations have been made during the last four years, especially in France, and this lymphocytosis has been found in nearly all cases of tabes (in 121 out of 126 cases recorded). *See* p. 93.

Complications.—A most important complication is general paralysis of the insane. The association of the two diseases is not rare. Sometimes the symptoms of tabes appear first and those of general paralysis afterwards, in other cases those of the latter disease first develop.

Hemiplegia from cerebral hæmorrhage or thrombosis or from cerebral syphilis is a rare complication.

Syphilitic diseases of the brain (gummata, syphilitic meningitis, spinal syphilis, and the various forms of cerebro-spinal syphilis) are also rare complications. These syphilitic affections are often relieved by anti-syphilitic treatment, whilst the true tabetic symptoms remain unaltered. Tachycardia and glycosuria are very rare complications.

Valvular diseases of the heart, especially aortic disease, are not uncommon. Leser found that amongst 96 cases of tabes, 18 had aneurism of the aorta. Arullani found signs of aortic diseases (aortitis, regurgitation, or aneurism) in 40 out of the 68 cases of tabes which he examined, the most common form being aortic regurgitation. Aortic aneurism is pro-

FIG. 138.—Tabes Dorsalis. Charcot's disease of ankle joint. Swollen ankle diseased; smaller ankle normal.

bably often an indirect result of syphilis, and both aortic diseases and tabes when occurring in the same individual may be regarded as due to previous syphilitic infection. A history of syphilis was obtained in 77 per cent. of the cases recorded by Arullani.

Forms of the Disease.—(1) In the common form of tabes the legs are chiefly affected—*tabes inferior*. The symptoms are those already described.

(2) In a rare form of tabes—*cervical tabes* or *tabes superior*—the upper limbs are chiefly affected. Shooting pains, other sensory symptoms and ataxia are present in the arms, whilst the legs are not affected, or only slightly affected. The deep reflexes of the arms are lost. The knee-jerks are present at first, but disappear later. Often gastric crises occur early. The pupils are usually affected (inequality and Argyll-Robertson pupils). The cervical form of tabes is very rare (1 case in 106—Dejérine). The prognosis is more unfavourable than in other forms, and the cranial nerves are more liable to be affected.

(3) Onset with *optic atrophy*. This form has already been mentioned.

The patient first seeks medical advice on account of failure of vision. Optic atrophy may be the only symptom of the disease at first; often for a long period very few other symptoms are present. But in these cases examination usually reveals absence of the knee-jerks and of the tendo Achillis reflexes; on inquiry a history of shooting pains may be often obtained, though these may have been very slight; and the only other symptoms may be muscular hypotonus. It is in these cases in which optic atrophy develops at the onset, and in which very few symptoms are present, that muscular hypotonus is a valuable confirmatory sign of the disease.

In the group of cases of tabes with early optic atrophy, the spinal cord symptoms are usually (but not invariably) mild and late in their development. It may be many years before ataxia develops. When the patient becomes blind often, but not always, the cord symptoms cease to progress. If optic atrophy occurs in tabes it is generally a very early symptom, but in some cases it develops after the other symptoms of the disease have become well marked, and in these cases the spinal symptoms may cease to progress when the optic atrophy appears.

(4) Sir William Gowers has described a form of tabes in which neuralgia is the most prominent feature—*tabetic neuralgia.* The knee-jerks are not lost, and there is no ataxia. He regards the disease as the result of a peculiar kind of tabetic toxin. The pains present many of the variations met with in ordinary tabes, and are usually severe; they are greatly increased by damp cold. Often a correct diagnosis is not made at first. A careful examination for the early signs of tabes is necessary, also the nature of the pain is important. Many causes produce pains resembling the deep dull prolonged pains of tabes; but the real " lightning pains," momentary stabbing the skin or flashing along it, and varying in seat are " hardly ever caused by any other morbid process " (Gowers).

(5) *Juvenile tabes—hereditary syphilitic tabes.* Tabes dorsalis is exceedingly rare under the age of twenty, but a number of cases are on record in medical literature, in which the disease commenced in childhood or youth. Such cases have been described as infantile or juvenile tabes, or hereditary syphilitic tabes; and though they are extremely rare, the number on record is steadily increasing. The reports of the cases published show that usually the parents have suffered from syphilis, or the tabetic children have presented signs or symptoms of hereditary syphilis; in other cases of juvenile tabes the patient has been infected with syphilis at an early period of life, often through a syphilitic nurse (extra-genital syphilitic infection). In some cases one of the parents of the young tabetic patient has suffered from both syphilis and tabes, or from general paralysis of the insane, or from cerebral syphilis (*see* p. 294).

Optic atrophy has been a common symptom in juvenile tabes; bladder

symptoms (paresis of the bladder, incontinence and retention of urine) have also been frequently present at an early period of the disease. In juvenile tabes females are affected more frequently than is the case in tabes of the adult. In 50 cases on record, 28 were females and 22 males.

The clinical history in three of my cases may be summarised as follows:

First case, girl aged eight; signs of early tabes; father had suffered from syphilis, and now presents the symptoms of advanced tabes; mother has had four miscarriages.

Second case, boy aged thirteen; signs of early tabes; teeth characteristic of congenital syphilis (Hutchinson's teeth).

Third case, girl aged seventeen; symptoms and signs of tabes for seven years; father has had syphilis; mother has had several miscarriages.

All three patients were blind, owing to primary optic atrophy. In two cases there was evidence of congenital syphilis; in the third case, congenital syphilis was probable, the father having suffered from syphilis.

Tabes in women. Mendel estimates that sterility is nearly three times more common amongst women suffering from tabes than amongst non-tabetic women in the same social position. In females, tabes runs a more chronic and a milder course than in men. The ataxic stage develops later; but Mendel thinks that complete amaurosis is more common in female than in male tabetic patients.

The **course** of tabes is usually very chronic. The duration may be twenty years or longer; in rare cases, however, death occurs after a few years. The disease is progressive, but the symptoms may remain stationary in the first stage for a long period, even for ten or twenty years. Also many cases show little progressive tendency, and in some the disease becomes practically arrested. In the cases commencing with optic atrophy the course is, as a rule, unusually long, and very many years may elapse before ataxia develops. Even when the third stage of tabes is reached the patient may live for years. Death occurs from some complication, such as cystitis, pyelonephritis, or other kidney complications, bed-sores, pyæmia, or from intercurrent diseases such as pneumonia, phthisis or bronchitis. When general paralysis is a complication, death may be due to this affection.

Marie and Moquot have published statistics of 66 cases of tabes with respect to the age at which death occurred—34 (i.e. 51·5 per cent.) were over 60 years of age at death, 55 (i.e. 83·3 per cent.) were over 50. When death occurred early the cause was not tabes, but some intercurrent disease. Of the 66 cases, 20 were blind. From these and other statistics it is evident that though tabes causes impairment of health and much suffering, it frequently does not directly shorten life.

The **prognosis** is usually unfavourable; but, as already mentioned, the disease not infrequently becomes arrested, especially in the first stage, and the subjective symptoms disappear entirely and the patient feels well; though signs such as the absent knee-jerks and the Argyll-Robertson pupil remain. In such cases, when death has occurred from some complicating

disease and an autopsy has been obtained, the pathological changes characteristic of tabes have still been found in the posterior columns.

It is only when the disease has followed soon after syphilis, or when its development is rapid, that anti-syphilitic treatment is likely to do good. In spite of the persistence of attacks of shooting pain, the disease does not always advance. In cases in which optic atrophy occurs the prognosis is usually good as regards the spinal symptoms; the patients generally remain for a long period in the first stage, and if ataxia should develop it is usually very late. To this general rule there are exceptions. Unfortunately the optic atrophy leads to complete blindness in most cases, but it is not invariably progressive.

Paralysis of ocular muscles and even ophthalmoplegia may disappear. Bladder symptoms are often temporary, and gastric crises may cease in course of time.

If attacks of pain or gastric crises are very frequent and severe, the patient is soon unable to follow his employment. In other cases, however, the tabetic symptoms do not cause much interference with work, and the patient may be able to follow some occupation for years.

When ataxic and bladder symptoms occur early, a more rapid course of the disease may be expected.

When symptoms of general paralysis of the insane are associated with those of tabes, the prognosis is very grave, and is that of the mental disease.

Diagnosis.—The difficulty in diagnosis is chiefly at the early pre-ataxic period. Shooting pains often suggest early tabes, and further examination or the development of the case confirms this suspicion. The sharp, shooting, stabbing or momentary pains (described on p. 304), each of a very short duration, but repeated in paroxysms, are almost characteristic of tabes.[1] The other forms of tabetic pains are not characteristic. But the diagnosis should not be based upon one symptom alone.

There are three symptoms which, when all present, render the diagnosis of early tabes justifiable—(1) shooting pains of the typical character; (2) absent knee-jerks or tendo Achillis jerks; (3) the Argyll-Robertson pupil. Even when only two of these three symptoms are present the diagnosis is very probable.

It is important to remember that the first symptoms which attract the patient's attention may be attacks of vomiting (gastric crises), or failure of vision from optic atrophy, or double vision from paresis of ocular muscles, or slight bladder troubles, or painless joint disease (Charcot's joint disease). The development of any of these symptoms should always rouse the suspicion of tabes, and should lead to a careful examination for other signs of this disease. The combination of primary optic atrophy or gastric crises with loss of the knee-jerks renders the diagnosis of tabes exceedingly probable, and if there be a history of shooting pains also the diagnosis is practically certain.

[1] I have occasionally obtained a description of similar pains in diabetes mellitus.

The absence of the knee-jerks is of the greatest diagnostic importance, since they are probably never absent in health. But before concluding that the knee-jerks are absent it is always desirable to try the method of examination recommended by Jendrássik and Buzzard (*see* p. 64). Absence of the knee-jerks is a very early sign of tabes, and the knee-jerks are almost always absent in this disease. But in rare cases the knee-jerks are present at first, though other symptoms point to tabes.

The tendo Achillis reflexes are usually absent in the very rare cases in which the knee-jerks are present. Hence the loss of the former reflex is of diagnostic value.

The Argyll-Robertson pupil is one of the most valuable sign of tabes; but it is not invariably present. (I have found it absent though I have been able to confirm the diagnosis of tabes by pathological examination of the spinal cord.)

Lymphocytosis of the cerebro-spinal fluid is nearly always present.

The following signs and symptoms when present are confirmatory evidence of tabes—irregular shape of the pupils (not quite circular, eccentric); zones of diminished cutaneous tactile sensation in the trunk;[1] muscular hypotonus (leg when extended being raised to 90° or more from the horizontal level); analgesia of the calf muscles (on the deepest pressure); and loss of the vibrating sensation on the feet or legs.

The detection of the earliest indication of ataxia is of great importance (*see* p. 298); but the diagnosis can usually be made in the pre-ataxic stage.

Failure of vision owing to primary optic atrophy may be a very early symptom of tabes and the first for which the patient seeks advice. For a long time few other symptoms may be present. In all cases of primary optic atrophy, therefore, careful examination should be made for signs of tabes.

The form of tabes beginning with optic atrophy has already been described. The other symptoms may be simply loss of knee-jerks and loss of tendo Achillis reflexes. Shooting pains may also be present, but often they are slight. Usually there is the Argyll-Robertson pupil. Muscular hypotonus is a very useful diagnostic sign in favour of tabes in these cases; and occasionally there are zones of impaired tactile sensation on the chest and loss of the bone sensibility (vibrating sensation) in the legs. Signs of early ataxia are important, but they are usually absent in these cases for a long period.

The diagnosis from other affections causing primary optic atrophy (i.e. disseminated sclerosis, general paralysis of the insane, basal syphilitic meningitis, tumour pressing on the optic chiasma) has to be carefully considered. An important point with respect to the visual defect caused by the primary optic atrophy in tabes is the fact that central scotomata do not usually occur. The presence of a central scotoma in a case of optic atrophy is evidence against tabes; whilst in the optic atrophy of

[1] Often absent in very early tabes in the pre-ataxic stage.

disseminated sclerosis and the basal syphilitic meningitis central scotomata often occur.

From *peripheral neuritis* the diagnosis is usually easy. In an uncomplicated case of tabes there is no actual paralysis, at least until the third stage of the disease; but in peripheral neuritis weakness and paresis of the legs, dropped feet (and sometimes dropped wrists), occur in a comparatively short time after the onset of the symptoms. The chief difficulty is in the diagnosis of the rare cases of neuritis in which ataxia is the prominent symptom—neuritic pseudo-tabes. But even in this form of neuritis some weakness of muscles can be usually detected; also changes in the electrical reactions (partial or complete reaction of degeneration) may be present; muscular tenderness is usually a prominent symptom;[1] the pains in the legs are of a dull aching character and not of the sharp stabbing kind met with in tabes; the pupils nearly always react to light; and there are no girdle pains and no zones of diminished tactile sensation on the chest. Also optic atrophy does not occur in peripheral neuritis. Further, in the neuritic cases there is usually a distinct history of alcoholism, arsenical poisoning, or of one of the causes of neuritis. In peripheral neuritis the symptoms reach their height in a few weeks or months, and then either the disease ends fatally or there is slow recovery.

In *diphtheritic paralysis* there is the history of diphtheria or of a suspicious throat affection; the course is more rapid; there are no sharp stabbing pains; paralysis of accommodation and of the soft palate, difficulty of swallowing, or paralysis of the laryngeal muscles may precede the neuritic ataxia or paralysis. The development of actual paralysis is evidence against the diagnosis of uncomplicated tabes.

In *ataxic paraplegia* the knee-jerks are increased, the legs become spastic, ankle-clonus and the extensor form of plantar reflex are often present. There are no ocular symptoms, and usually there is no anæsthesia.

Cerebellar tumour often causes ataxia; but in this affection optic neuritis and headache are usually present, and there are no shooting pains or sensory disturbances in the legs.

In severe forms of *diabetes mellitus* the knee-jerks and tendo Achillis jerks are often absent, and there are frequently pains in the legs; perforation ulcers are in rare cases present on the feet, and impotence often occurs; so that occasionally the question of tabes arises. In diabetes, however, the pains are usually dull and aching, rarely sharp and shooting, the Argyll-Robertson pupils are absent, there is no girdle sensation, and the urine is characteristic of diabetes. In all cases in which a perforating ulcer is present and the knee-jerks absent both diabetes mellitus and tabes should be considered, and the urine always examined for sugar and other changes.

[1] Muscular hyperalgesia—great pain on pressure over the muscles of the calf or arm—is usually a prominent symptom in peripheral neuritis. In many cases of tabes no pain can be produced by very strong pressure of the calf muscles.

A *lumbar myelitis* often causes loss of knee-jerks ; but here paralysis of the muscles of the legs with wasting, paralysis of the bladder and a sacral bed-sore, usually indicate the diagnosis.

In *disseminated sclerosis* ataxia may occur, but the knee-jerks are usually present, there is often tremor of the hands of the characteristic type (intention tremor) with nystagmus and scanning speech, whilst typical shooting pains and the Argyll-Robertson pupil are absent. The Babinski extensor type of plantar reflex, ankle-clonus and spastic condition of the legs are signs in favour of disseminated sclerosis, and do not occur in uncomplicated tabes ; also if optic atrophy be present, central scotomata would be in favour of disseminated sclerosis and against tabes.

For the diagnosis from Friedreich's disease, *see* p. 350.

General paralysis of the insane may cause loss of knee-jerks, Argyll-Robertson pupils, and in rare cases optic atrophy. Also tabes and general paralysis are not infrequently associated. Indications of general paralysis would be tremor of the lips and tongue, the peculiar speech affection and mental symptoms.

The diagnosis of tabes from syphilitic pseudo-tabes is considered on p. 398.

From the cases described by v. Leyden as "*acute ataxia*" (usually due to a myelo-encephalitis), the diagnosis is easy. In tabes the development of ataxia is very gradual ; in acute ataxia rapid ; though there are rare cases of tabes in which the ataxia develops somewhat rapidly. The cases of acute ataxia (myelo-encephalitis) usually follow some acute ailment or infectious disease ; the course is not progressive ; before the ataxia there are no other well-marked nervous symptoms ; the Argyll-Robertson pupil is absent, and the knee-jerks are present.

In recent years much attention has been directed, especially in France, to the examination of the cerebro-spinal fluid in tabes. It has been demonstrated that in tabes there is almost always an excess of lymphocytes in the fluid obtained by lumbar puncture —lymphocytosis.

Hence the examination of the puncture fluid may be an aid to diagnosis; but it should only be resorted to in exceptional cases (*see* p. 93).

Morbid Anatomy and Pathology

Microscopical examination has shown that in tabes the most important change is a degeneration of the sensory neurons of the spinal cord, in which the lesion affects chiefly the central axon, i.e. the long process of the neuron within the posterior columns of the spinal cord.

As already described, the posterior columns of the cord consist of exogenous and endogenous fibres. The exogenous fibres arise from cells of the posterior root ganglion—outside the spinal cord. The endogenous fibres arise from nerve cells within the spinal cord. It is the former fibres which suffer first and chiefly in tabes. Moreover, certain of these fibres of the posterior root within the cord suffer before the others, i.e.

the toxin causing the degeneration has an *elective* action on the posterior root fibres.

The changes in the upper part of the cord are chiefly of the nature of a secondary ascending degeneration similar to the ascending degeneration caused by non-tabetic lesions.

FIG. 139. — Spinal Cord; advanced tabes dorsalis. Weigert's stain. Pale area in posterior columns degenerated. Lowest section, lumbar region ; posterior columns degenerated with exception of posterior ventral fields (close to grey commissure). Middle section, dorsal region. Uppermost section, cervical region ; degeneration in Goll's columns only.

I. **Advanced Changes.**—In advanced cases of tabes the posterior columns of the cord appear grey and somewhat translucent to the naked eye. They are often small and shrunken ; hence the shape of the transverse section of the cord is altered. When sections of the spinal cord are stained with carmine or aniline blue-black the posterior columns are more deeply stained; when stained according to Weigert's or Pal's method they are less deeply stained than the rest of the white matter. Microscopic examination reveals sclerosis of the posterior columns, the exact distribution of which will be subsequently described. The sclerosed tracts present increase of neuroglia connective tissue and degeneration, partial or complete, of nerve fibres. On examination of parts at which the degeneration is not complete some fibres are seen to be swollen and their medullary sheath distended ; other fibres present granular degeneration of both axis cylinder and medullary sheath ; and at an advanced stage of this change only granular material is seen at the positions previously occupied by nerve fibres. Many nerve fibres present only the changes of simple atrophy (*see* p. 40).

When the degeneration is complete, empty spaces may be seen in the neuroglia, from which all traces of the nerve fibres have disappeared.

The neuroglia connective tissue is often increased, and may fill the spaces left by the degenerated fibres. In the neuroglia spider cells may be found and the walls of the blood vessels are sometimes thickened. Compound granular cells are usually absent, but in rare sub-acute cases they may be seen in the neuroglia.

There can be little doubt that the changes are due to primary parenchymatous degeneration of nerve fibres, and that the excess of neuroglia connective tissue is a secondary change, and not the result of primary chronic inflammation. Weigert has shown that when one tissue element of a complex structure undergoes atrophy, the other has a tendency to undergo hypertrophy or to proliferate.

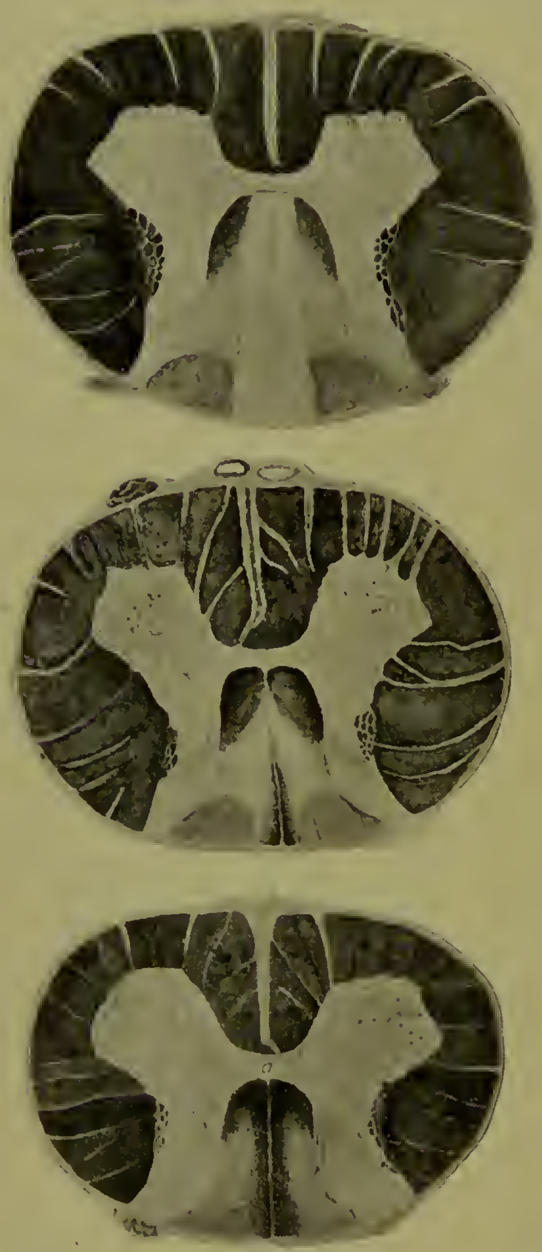

Fig. 140.—Section of Spinal Cord: early tabes. Weigert's stain. Pale areas in posterior columns = degenerated parts. (Case recorded by writer in *Medical Chronicle*, October, 1897.) The highest figure = lower cervical region; middle figure = upper lumbar region; lowest figure = lower sacral region.

The changes in the posterior columns of the spinal cord are usually most marked in the lumbar and lower dorsal regions; above this part the degeneration in the posterior columns gradually diminishes in most cases, and becomes localised to the columns of Goll, having thus the localisation of the ascending degeneration seen after transverse lesion of the lower part of the cord.

In an *advanced* case of tabes, in the lumbar region there is degeneration of the whole of Burdach's and Goll's columns, with the exception of two regions. The regions usually spared are (i.) the ventral posterior fields, just adjacent to the posterior commissure, and (ii.) the dorso-medial bundles (centrum ovale of Flechsig), two small bundles of fibres in Goll's columns, one on each side of the posterior median fissure, forming together an oval-shaped area. The lower sacral region is affected later than the lumbar, and a small patch of degeneration is present close to the median part of the posterior horns, whilst the rest of the posterior columns is unaffected.

In most cases in the upper dorsal region, the degeneration is situated more and more towards the median septum, until finally only the columns of Goll are degenerated. If the upper dorsal posterior root fibres are also affected, then more complicated areas of degeneration are produced

in the dorsal posterior columns, especially if diseased spinal segments are separated by healthy segments. The columns of Goll are then degenerated, and also there is a separate zone of degeneration in Burdach's column on each side, close to the posterior horn or separated a little distance from the horn, according to the level of the section above the degenerated roots. If all of the posterior root fibres of the sacral, lumbar, and lower dorsal regions are diseased, then almost the whole of Burdach's and Goll's columns is degenerated in the mid-dorsal region, as in the lumbar region.

In the cervical region the columns of Goll alone are degenerated, if

Anterior.

Median part.

External part.

Posterior.

FIG. 141.—Section of Dorsal Region of Spinal Cord in Tabes. Weigert's stain; high power of microscope. The grey matter with its normal fine nerve fibres is seen running through the middle of the figure. Exactly in the centre of the figure is a roundish area in the grey matter, in which the fine nerve fibres are absent. This is the column of Clarke, in which the nerve fibres have degenerated.

only the posterior root fibres of the lower part of the cord are affected (as is very often the case in tabes). When the cervical root fibres are affected, in addition to the dorsal and lumbar fibres, then there is an area of degeneration in the cervical posterior external column, close to the posterior horn; also the columns of Goll are degenerated, and between the two areas there are degenerated fibres which have come from the dorsal nerve roots. When all of the cervical root fibres are affected, in addition to the degeneration of dorsal and lumbar fibres, then almost the whole of the cervical posterior columns is degenerated.

In cases of cervical tabes, when only the cervical root fibres are affected, at an early stage, Goll's columns may be free, and the degeneration found is close to the posterior horns of grey matter. But at a later stage the dorsal nerve roots usually become affected also, and then degeneration is found in addition in Goll's columns in the cervical region.

The changes in the cord in tabes, as already mentioned, are due to

degeneration of the fibres in the posterior columns which come from posterior nerve roots. But fibres from the posterior roots also enter the grey substance after passing through the outer part of the posterior columns; and others enter the posterior horn directly. In the normal cord the latter form a bundle of fine fibres at the tip of the posterior horn—Lissauer's column. In advanced tabes this column of fibres and the fibres passing from the posterior columns into the posterior grey horns may be degenerated. The fine nerve fibres of Clarke's columns in the grey matter are degenerated, but the cells of these columns

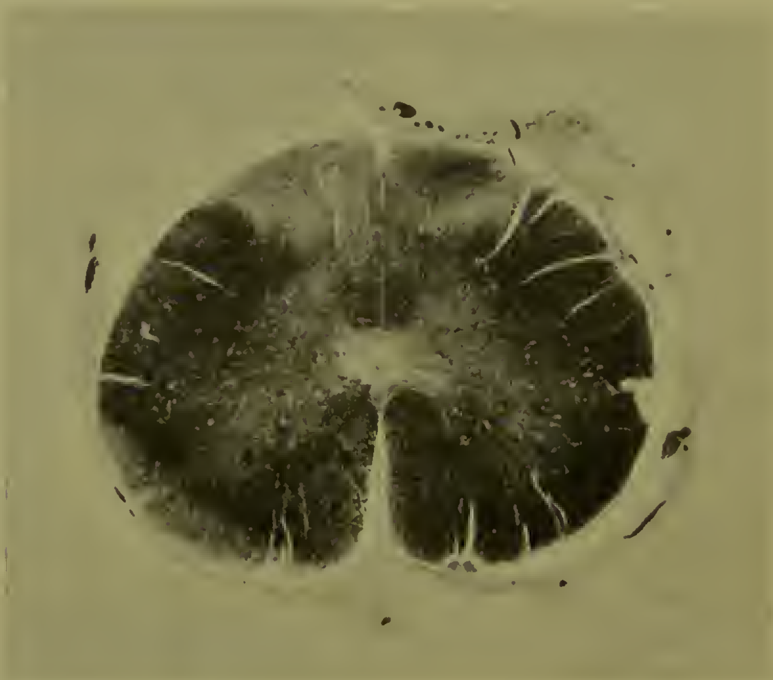

Fig. 142.—Spinal Cord. Early tabes dorsalis. Lower lumbar region. Weigert's stain. Note pale area of degeneration in posterior root zone (close to posterior horns). To the median side of this degeneration, i.e. at a small area on each side of the posterior median septum, and also close to the posterior commissure, the structure appears normal.

usually are normal. In very advanced cases the cells of Clarke's columns may be degenerated, and in such cases the fibres of the direct cerebellar tracts are also degenerated. In rare cases of advanced tabes there is degeneration of the fibres of the antero-lateral ascending tract and very rarely the crossed pyramidal tracts are slightly affected. In the median grey matter and in the anterior horns the reflex collateral fibres are often degenerated. The nerve cells of the posterior horns may be diminished in number and atrophied and present the changes of chromatolysis. The extra-medullary part of the posterior root is atrophied and degenerated in the lumbar and lower dorsal regions in advanced cases of tabes.

II. **Early Changes.**—The degeneration of the posterior columns begins in the middle and lower lumbar or upper sacral regions; and when the examination of the cord is made at an early stage of the disease, there is an area of degeneration in Burdach's column in the lumbar region

close to the middle third of each posterior horn of grey matter—posterior root zone. From this point the degeneration extends towards the middle line. This degeneration in the posterior external columns is present in almost all cases in the lumbar and lower dorsal regions, whilst it is often absent at higher parts, and in the upper dorsal and cervical regions there is simply ascending degeneration in the columns of Goll.

The form and size of the areas of degeneration vary somewhat in different cases. Sometimes the degenerated zone in Burdach's column is close to the posterior horn, sometimes a little to the median side. In cases a little more advanced, the degeneration on each side extends towards the middle line and the two areas meet. When the sacral region is diseased, then the degeneration in the lumbar region extends to the middle line (*see* Fig. 143). In early tabes the whole of the posterior

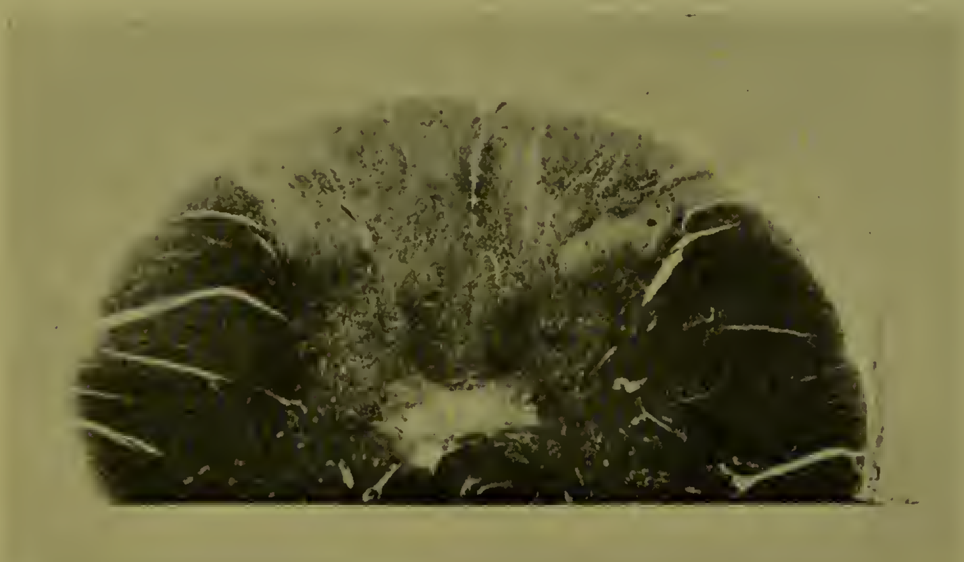

FIG. 143.—Spinal Cord: lumbar region. Early tabes dorsalis. Weigert's stain. Posterior columns and horns shown in photograph. Note posterior root zone degenerated (pale): posterior ventral field and Flechsig's oval field (close to posterior median septum) not degenerated: posterior external field less affected than posterior root zone.

external column is not degenerated in the lumbar region; often the posterior external field is little affected, and is sharply defined from the adjacent degeneration; also the dorso-medial bundle and the ventral posterior field are spared. The middle diagram in Fig. 140 shows the shape of the degenerated area in the lumbar region in a case of early tabes.[1] In some early cases Lissauer's column is not affected, in others only slightly degenerated.

(The posterior external field, which is little affected, also contains fibres from posterior roots, and might, therefore, be expected to be much diseased.)

By studying the development of the spinal cord in the embryo at different stages, with the aid of Weigert's method of staining, Trepinski

[1] Recorded by the writer in the *Medical Chronicle*, Oct. 1897.

has mapped out several areas in the posterior columns. These areas can be separated, because the nerve fibres composing them acquire their medullary sheath at different times. Trepinski has shown that the exact localisation of the degeneration in the posterior columns in tabes coincides with the extent of definite embryological areas of fibres. Thus in an early case of tabes the area of degeneration in the posterior columns (as shown in the middle diagram, Fig. 140) corresponds in its localisation to the third system of fibres differentiated embryologically by Trepinski. In other words, the portion of the posterior columns degenerated at an early stage of tabes corresponds to a bundle of fibres which acquire their medullary sheaths at a different period from the rest of the fibres in the posterior columns. In more advanced cases of tabes the degeneration extends further to the other embryological fibre systems.

FIG. 144.—Posterior Columns and Posterior Grey Matter of one-half of the Spinal Cord. Early tabes dorsalis. Weigert's stain. Pale area in posterior columns degenerated.

Hence in tabes the lesion in the cord is one affecting at first only certain fibres coming from the posterior nerve roots—fibres which correspond to a definite embryological section of the posterior columns; but the changes are progressive, and finally all the fibres of the posterior roots may be involved within the cord.

The pia mater over the posterior columns is thickened and well nucleated in advanced cases. but in recent cases no definite change may be detected. The walls of the blood vessels of the posterior columns are thickened in the sclerosed parts, and occasionally they are hyaline; but in early cases no definite change may be detected.

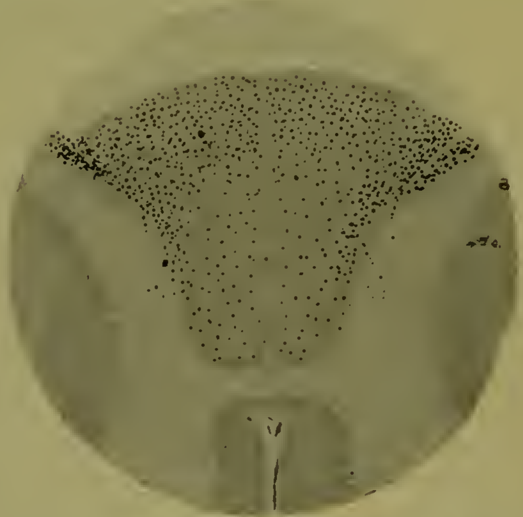

FIG. 145.—Early Tabes Dorsalis. Marchi's stain. Degenerated fibres = black dots. Note degenerated fibres most numerous close to posterior horn of grey matter.

III. **Posterior Roots.**—As already stated the posterior nerve roots are degenerated in advanced cases. There is atrophy and disappearance of nerve fibre with increase of connective tissue. The degeneration can often be traced up to the spinal ganglion of the posterior root, but on reaching the ganglion it ceases, or it may extend slightly to the peripheral

side thereof. In more recent cases perineural small cell infiltration may be sometimes detected, or there may be perineural fibrous thickening at the part of the posterior root which is surrounded by the dural sheath.

But in cases of tabes in which the pathological examination is made at an early stage of the disease, the posterior nerve roots may appear normal or present only very slight and doubtful changes; and yet the posterior root fibres within the cord may be markedly degenerated, as in a case recorded by the writer (*see* Fig. 147).

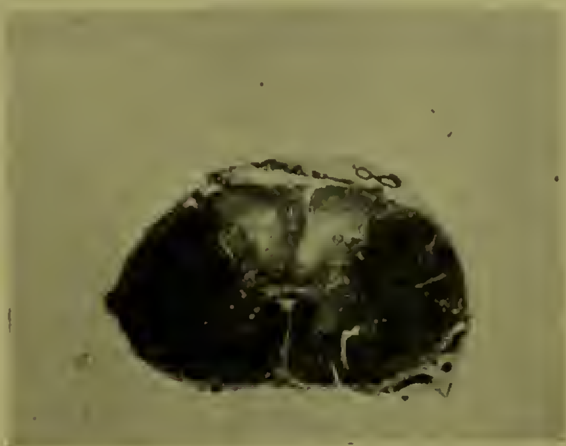

Fig. 146.—Cervical Tabes Dorsalis: cervical region of Spinal Cord. Arms markedly affected. Note marked degeneration in Burdach's columns (pale area). Pal's stain.

IV. **Spinal Ganglia.**—The condition of the *cells of the spinal ganglia* of the posterior nerve roots has varied. In some cases there has been atrophy of these cells; in other cases only slight changes have been detected. Marinesco, Schaffer, Maragliano and others have found no essential changes in the cells. Several observers regard the changes in the cells which have been detected in some cases, as simply secondary to the degeneration of the posterior nerve root fibres. It is worthy of note that in early tabes, though the changes in the spinal cord may be well marked, both the posterior nerve roots and the posterior ganglia may appear normal or almost normal, and the decided degeneration commences at the point where the posterior root fibres pass through the spinal pia mater.

V. **Peripheral Nerves.**—The peripheral sensory nerve fibres often present changes, which are seen chiefly in the small sensory nerves in the skin of the legs. The degeneration of these nerves has been found in portions of skin removed during life. The changes diminish as the larger nerves are reached, and they are not proportionate to the spinal degeneration. In advanced cases of tabes the changes in the peripheral nerves are much less than those in the posterior nerve root fibres within the cord.

When tabes has been complicated with peripheral neuritis decided changes have been found in mixed motor and sensory nerves.

VI. **Cranial Nerves and Medulla.**—In the medulla the spinal trigeminal nerve root, the solitary bundle, and the posterior vagus nucleus are sometimes degenerated. The nucleus ambiguus is said to be never affected.

In rare cases the nerve cells of the sensory trigeminal nuclei, the nuclei of the hypoglossal and of the spinal accessory nerves, and the nuclei of nerves supplying the ocular muscles have been degenerated;

but the motor nerves to the eye muscles, as well as the vagus and recurrent laryngeal and hypoglossal nerves, may be degenerated without the occurrence of any change in the nuclei of these nerves.

Very rarely there is degeneration in the auditory nerve. Oppenheim has recorded degeneration of the cells and nerve fibres of the Gasserian ganglion.

Of the cranial nerves the optic are most frequently affected. The degeneration of the optic nerves (optic atrophy) apparently begins at the periphery and extends towards the brain, at least on pathological examination the nerve near the eye is markedly affected, whilst in the chiasma and optic tracts the changes are less marked. Early degeneration of the ganglion cells of the retina has been described, and the optic nerve atrophy has been regarded as secondary to this change; but in other cases the retinal ganglion cells have not been degenerated, although blindness has been present for many years.

Numerous observations have shown that the tabetic optic atrophy is a simple atrophy. The tabetic optic nerve atrophy is regarded by some as the result of a constriction or compression of the nerve, in its intra-ocular portion, by neuroglia proliferation, or vascular sclerosis.

It is interesting to note that, at their entrance into the eye-ball, the optic nerve fibres lose the medullary sheath, and only the axis cylinders are prolonged into the retina. May it not be that at this point, where the nerve fibres lose their medullary sheaths, they are more liable to be affected by toxins circulating in the blood?

Based on the examination of the optic nerve in twenty-one cases of tabes with amaurosis, André Léri concludes that the lesion in tabetic amaurosis is an interstitial neuritis, a syphilitic cirrhosis of vascular origin and a syphilitic meningitis. The meninges are infiltrated markedly with lymphocytes. In the nerve itself there is a marked new formation of vessels and interstitial tissue or neuroglia. The vessels gradually become sclerosed and obliterated, and the nerve is deprived of its blood and lymph circulation. As a result the nerve fibres atrophy. Léri concludes that the optic atrophy does not begin in the retina, and that it is *not* due to a primary elective degeneration of the cells of origin of the fibres of the optic nerve (i.e. multipolar cells of the retina).

Weigert has drawn attention to neuroglia proliferation in localised patches in the molecular layer of the cerebellum.

VII. **Origin of Pathological Changes.**—As already stated the characteristic lesion in tabes is degeneration of the intra-medullary fibres of the posterior nerve roots—i.e. the posterior root fibres in their course within the spinal cord (*exogenous* fibres). In advanced cases other fibres in the cord—endogenous fibres, or fibres not derived from the posterior roots—become finally degenerated from the irritation set up by the adjacent sclerosis and vascular nutritional changes. Obersteiner thinks that it has only been definitely proved that the intra-

medullary continuations of the posterior roots are at first degenerated in tabes.

With respect to the exact pathology of tabes, an important question for consideration is whether the degeneration of the posterior root fibres within the cord is *secondary to some process outside the cord*—i.e. *whether it is due to a lesion affecting the sensory neuron at some part outside the cord.* There are several views as to the point of origin of the changes.

Changes in the posterior roots outside the cord are distinct in advanced cases. In cases of medium severity, not infrequently the disease of fibres in the posterior roots outside the cord is much less advanced than within the posterior columns. In early tabes, the posterior nerve root (between the ganglion and the surface of the cord) may appear normal, or almost normal, whilst in the posterior columns of the cord the posterior root fibres are markedly degenerated. Hence the posterior surface of the cord is the point at which the changes in the posterior root fibres either commence or at least become well marked.

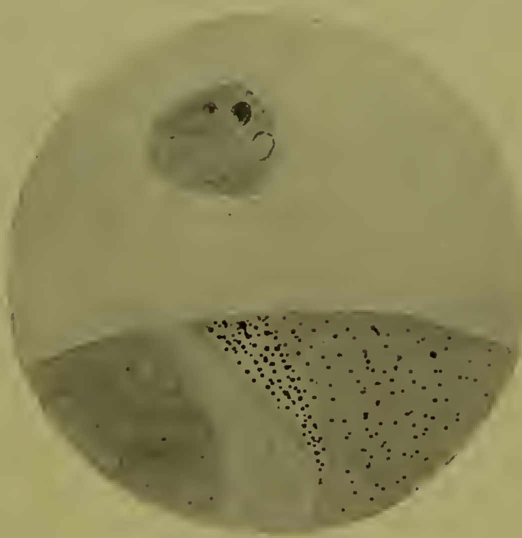

Fig. 147.—Early Tabes Dorsalis. Posterior nerve root in transverse section, close to surface of Spinal Cord. Marchi's stain. Note black dots, degenerated nerve fibres, in posterior columns of Cord, shown at lower part of figure. These are most numerous close to posterior horn, near middle line of figure. In the posterior root no degenerated fibres can be seen.

Thickening of the pia mater (chronic meningitis) may be found in tabes; it has been regarded as the cause of the changes in the posterior root fibres, and the lymphocytosis of the cerebro-spinal fluid is in favour of this view. But chronic meningitis is not found in all cases, and in the early stages of tabes the meninges often appear normal. Moreover, ascending degeneration of posterior root fibres within the cord is very rare in cases of spinal meningitis from other causes. In marked syphilitic meningitis the changes in the posterior columns do not correspond to those of true tabes.

Schmaus points out that the regularity and symmetry of the cord changes in tabes do not correspond to a lesion usually so irregular as meningitis.

Nageotte has described cell infiltration of the dura mater and arachnoid at the point where the nerve roots pass through these membranes, and he attributes the degeneration in the posterior roots to the extension of these changes from the membranes to the nerve fibres at this region. He concludes that tabes is the result of this localised lesion of the

posterior roots between the dura mater and the ganglion; that the changes extend sometimes as far as the ganglion; and that the lesion is a transverse interstitial neuritis of the root which is due to a chronic syphilitic meningitis.

A case of tabes which I have recorded appears directly opposed to this view. This case was of three years' duration. There was marked degeneration of the intra-medullary fibres of the posterior root, whilst the extra-medullary fibres (outside the cord) were normal or almost normal. Had the changes commenced at the point where the nerve

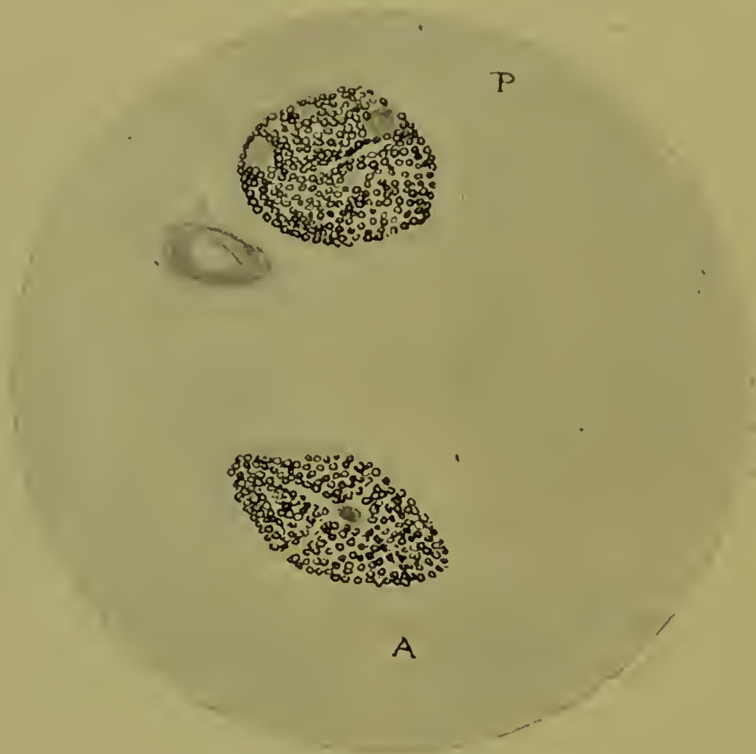

FIG. 148.—Anterior and Posterior Nerve Roots close to Spinal Cord. Early tabes dorsalis. Weigert's stain. Note posterior root (P) appears normal. A= normal anterior root.

passes through the arachnoid and dura mater (as Nageotte suggests) one would have expected definite change in the extra-medullary posterior root fibres close to the cord.

Obersteiner thinks that meningeal changes around the nerve roots are not sufficient to explain the degeneration in the latter. The anterior nerve roots would be liable to the same affection at this point, but usually they are not found to be diseased. Obersteiner has pointed out that the posterior root fibres in the normal cord are constricted as they pass through the pia mater and cortical sheath of the cord. At this region their medullary sheaths are markedly diminished and reduced to a minimum, or are absent. (This condition is clearly seen in sections stained according to Weigert's method.) Obersteiner and Redlich believe that this is a point of least resistance in the nerve fibres, and that an injury here—by a meningeal change, or

by pressure on the fibres through altered blood vessels lying close to them—is sufficient to cause the degeneration in tabes.

Obersteiner does not attach so much importance to a real meningitis as to a constriction in the pia mater (like that produced by the contraction of fibrous tissue in the liver in tertiary syphilitic disease). Also, the connective tissue layer on the surface of the cord may play a part in causing this degeneration. Obersteiner thinks that the contraction of the tissues just mentioned forms an anatomical basis predisposing to the development of tabes. In favour of this view are the facts that the intra-medullary part of the posterior root is more affected than the extra-medullary, or the former fibres only may be degenerated.

Orr and Rows regard tabes as "a system lesion, which begins as a parenchymatous degeneration of the sensory proto-neurons starting at the point where the neurilemma (or external sheath) is lost." This point being at the passage of the posterior root fibres through the pia mater. Here also the myelin sheath is greatly diminished and often appears absent. The loss of the neurilemma renders the intra-medullary portion of the sensory proto-neuron more open to the attacks of noxious agents at the point mentioned.

Degeneration of the posterior root fibres appears to commence at this point in certain cases of intra-cranial tumour, in some cases of diabetes mellitus and in several other diseases (*see* p. 37).

It is to be borne in mind, however, that tabes is not simply a disease of the spinal cord; the cerebral and spinal peripheral nerves, and especially the optic nerves, are often degenerated, and lesions are found in some cases in the nuclei in the medulla.

The *ganglia of the posterior nerve roots* have been regarded as the starting point of the changes by many writers. As the intra-medullary posterior root fibres and the fibres of the peripheral sensory nerves, which are often diseased also, are both connected with one cell, the lesion of tabes could be explained by a disease of these cells, i.e. those of the spinal ganglia (*see* Fig. 149).

But Marinesco, Maragliano and others have examined the spinal ganglia in cases of tabes, employing the most recent methods of staining, with practically negative results. They conclude that the various slight changes which they have found in the cells of these ganglia, present nothing characteristic, and that they cannot be regarded as the cause of the degeneration occurring in the posterior columns in tabes.

On the other hand Thomas and Hauser conclude from numerous observations, that the degeneration in the posterior root fibres is a simple atrophic process; the myelin sheath and axis-cylinders disappear slowly and progressively, and Wallerian degeneration is exceptional. They found slight changes frequently in the cells of the ganglia of the posterior nerve roots. These changes consisted chiefly in a process of slight atrophy and disintegration, which ends in the disappearance of the cell. They consider that it is difficult to appreciate the rôle which

such changes play in the pathogenesis of the atrophy of the posterior roots and of the spinal degeneration, but that they are too frequent, and too marked in certain cases, not to play some part in the pathogenesis of tabes.

The changes which have been found in the cells of the ganglia of the posterior nerve roots in tabes are, however, similar to those seen after division of the root in animals, and according to Köster probably they are of the nature of secondary changes due to the degeneration of the posterior root fibres within the cord. Köster has shown that division of the posterior roots in animals causes a reaction in the spinal ganglion cells. Some of the cells simply atrophy, others degenerate completely. Fibres connected with the latter degenerate and give a definite reaction with Marchi's stain between the ganglion and the surface of the cord; other fibres connected with the atrophied cells simply present an atrophied condition.

Marie formerly held the opinion that the first changes in tabes are situated in the spinal ganglia; but he allows that his view has not been confirmed by subsequent microscopical examinations. In all the cases which he has since examined, changes in the spinal ganglia could not be detected. Strong evidence against this theory is the fact that in advanced tabes degeneration may be found in those fibres which pass from the ganglion into the posterior root, whilst it is absent or very slight on the other side of the ganglion—i.e. in the fibres from ganglion cells to peripheral nerves. In advanced cases the central processes may be affected close to the ganglion cells, whilst the peripheral processes or fibres are not affected. Such a difference in the two processes from a nerve cell is not observed elsewhere in degeneration in the nervous system. This fact is also strong evidence against the view that the degeneration in nerve fibres is due to a functional change in the ganglion cells. Further, as Schmaus points out, it is strange that degeneration (in marked cases) can be sometimes traced in the posterior root fibres up to the posterior root ganglion, and yet the ganglion cells themselves may show no changes.

Marie and Guillain regard the changes in the fibres of the posterior columns as the result of a specific *lymphangitis of the posterior system of the lymphatics of the cord.* This system of lymphatics does not communicate with the lymphatics of the lateral and anterior columns. The lymph in the posterior columns ascends. The lymph spaces of the posterior pia mater communicate with those of the posterior columns. There is a common lymph system for the posterior roots, posterior columns and the posterior pia mater. The lymph is not contained in vessels, but passes along the intervals between the fibres and other structures. Homén has shown that lymph ascends to the cord in the peripheral nerves, and that toxins are conveyed more easily by the posterior roots than by the anterior roots.

Another view is that the change in tabes begins in the termination

of the sensory peripheral nerves, and that the cord lesion is secondary to these. v. Leyden and Dejérine have shown that degeneration is often present in the peripheral nerves; and, according to Oppenheim, the cutaneous sensory nerves (chiefly of the legs) are constantly degenerated. But these changes are not proportionate to the intensity of changes in the posterior root fibres, and it has not been shown that they occur before the cord changes. There is further difficulty in explaining how these changes in the peripheral nerves are transmitted to the cord. The spinal ganglion cells, as already described, often appear normal, or their lesions are slight. The posterior root fibres between the cord and spinal ganglia may be much degenerated in advanced tabes, but the fibres immediately on the peripheral side of the ganglia not degenerated; also, tabes usually begins at a definite level of the cord, since the loss of the knee-jerk is one of the earliest symptoms. But such a regularity and exact localisation has not been shown to occur in the peripheral degenerative neuritis of tabes. In marked peripheral neuritis from alcohol the cord changes are absent or slight, and yet the changes in the peripheral nerves are much more marked than in tabes. It must be allowed, however, this may be owing to the more rapid course in alcoholic neuritis, before degeneration has had time to occur in the cord.

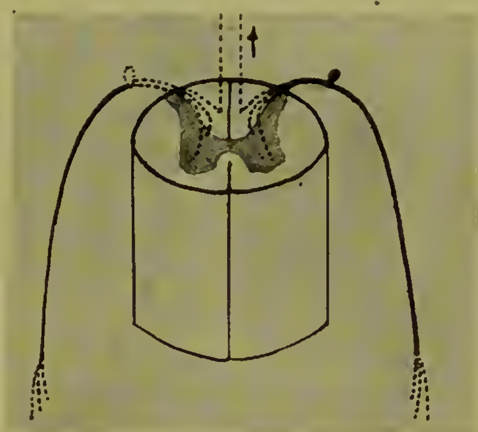

Fig. 149.—Diagram of Lesions of Posterior Root Fibres and Peripheral Nerves in Tabes Dorsalis. Fibres marked with dotted lines are degenerated. On right hand side of figure, early tabes, degeneration affects only fibres within the Cord, and in the peripheral nerve. On left-hand side, advanced lesion, the posterior root and its ganglion (outside the Cord) are also affected — indicated by dotted lines.

None of the views already mentioned are accepted by many writers, and the changes in the intra-medullary fibres of the posterior nerve roots, as well as those in the extra-medullary fibres of the roots and in the peripheral nerves, are all regarded as primary and due to the action of some toxin. It is interesting to note that the fibres of the posterior column are especially liable to degenerate in many diseases (*see* p. 376).

The present state of knowledge respecting the pathology of tabes may be summarized in the following statements:—

Tabes is a progressive disease of the sensory neurons of the cord, and often the sensory neurons of the brain are also affected.

It is not easy to decide where the first pathological change occurs in the sensory neuron.

The changes in the intra-medullary fibres of the posterior sensory neuron (spinal cord changes) are well marked; changes in the cells of the posterior ganglion are often slight or absent; those in the peri-

pheral fibres (peripheral nerves) are very slight and limited to the termination of these nerves. The changes in the peripheral nerve are disproportionate to those of the fibres within the cord. At the early stage, the changes in the intra-medullary fibres of the posterior nerve roots (fibres of posterior columns) are well marked; whilst the extra-medullary fibres of the posterior roots, just outside the pia mater, are normal or show only very slight changes. The changes in the posterior root fibres appear either to become most marked, or to begin where these fibres pass through the pia mater. Probably this is a point of diminished resistance of the fibres to toxic action. Possibly the comparatively feeble blood supply of the posterior columns of the cord has some influence in causing the degeneration to commence here.

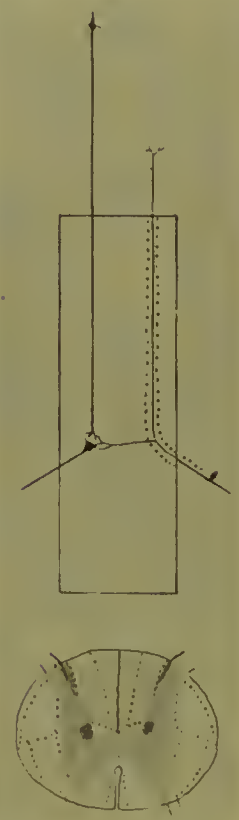

FIG. 150.—Tabes Dorsalis. Lower figure = transverse section of Spinal Cord, position of advanced lesions shaded — posterior columns (except posterior ventral field) and posterior vesicular column of Clarke. The upper figure = longitudinal section of cord. To the left are the anterior nerve root and its cell (lower motor neuron), and above it the upper motor neuron. To the right is the sensory neuron (posterior root, ascending fibres and collateral). The part degenerated is marked with double dots. The posterior root — external to the cord — is marked with single dots to indicate that it is degenerated in advanced case but not in early cases.

Within the posterior columns it is chiefly those fibres which are derived from the posterior root which degenerate, i.e. the exogenous fibres.

The degeneration of the posterior root fibres is elective. Certain fibres suffer more than others. Possibly the poison causing the degeneration of tabes has an elective action, affecting certain fibres chiefly, as in the case of lead poisoning, in which certain peripheral nerve fibres are chiefly affected.

The degeneration of tabes attacks a few neurons at first and then advances through the spread of the disease to other sensory neurons.

As a result of the degeneration of sensory neurons, at the lower part of the cord, ascending degeneration occurs in the posterior columns at a higher level.

Along with the degeneration of posterior root fibres within the cord, there is a proliferation of neuroglia connective tissue, sclerosis is produced and finally the proliferation of neuroglia extends to adjacent parts of the posterior columns which contain endogenous fibres.

The nature of the poison causing the degeneration of tabes is considered on p. 296.

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VIII. **Complications.**—In tabes there is a tendency to spontaneous fracture of bones. The calcareous salts of the bones have been found diminished (Regnard), and the Haversian canals dilated in scattered patches of bone. But these changes have not been present in all cases, and in some cases there has been a distinct sclerosis of bone tissue. The X ray photographs in some cases have shown a shadow of comparatively feeble intensity, attributed to rarefaction of bone.

The joint affections in tabes have received much attention, though the views as to their origin vary considerably. The joint surfaces are rapidly absorbed, and the ligaments destroyed. The synovial membrane is pale and villous ; the capsule thick and adherent to adjacent tissue, and in it calcareous deposits are sometimes seen. In marked cases the capsule may be atrophied and distended. The fluid in the joints is abundant ; it is usually bright or dark yellow in colour.

At the articular surfaces in some cases there is atrophy of cartilage and bone ; in others hypertrophy of these tissues. In the atrophic form the cartilage of the articular surfaces disappears and the bone is exposed. These exposed bony surfaces also gradually atrophy until the joint ending of the bone has disappeared. Proliferative changes and bony deposit may occur in the neighbourhood of the joint. The changes lead to dislocation and sub-luxations, and abnormal movements at the affected joints can be performed.

According to Rotter, analgesia of the joint surfaces is always present, and this condition, along with the inco-ordination of movements and the increased tendency to bone fracture, are important factors in the causation of the joint affection.

The relation of the joint affection to the changes in the cord has been much discussed. In recent years changes have been found in the peripheral nerves going to the joints and bones, and it appears probable, as Weizsäcker suggests, that the joint affection is largely due to these changes.

When actual paralysis has been present as a complication of tabes, it has been due to neuritis of peripheral nerves in some cases ; in other cases atrophy of the nerve cells of the anterior horns has been found.

When tabes is complicated by localised muscular atrophy the wasted muscles are yellowish in colour : some of the muscle fibres are degenerated markedly, have lost their striæ and present fine granules and nuclei within the sarcolemma ; some fibres are narrow but retain their striation ; whilst others are normal. Hence the reaction of degeneration is usually absent.

Condoleon has carefully investigated the pathology of these cases of muscular atrophy and found lesions in the cells of the anterior horns,

with very slight changes in the anterior roots and large nerve trunks, but with pronounced lesions in the intra-muscular nerves.

IX. **Cause of Symptoms.**—We have seen that the degeneration in tabes affects the sensory part of the nervous system, as is shown by the distribution of the pathological changes. It is a disease of the sensory neurons.

The changes in the posterior root fibres within the cord, and in the peripheral nerves, explain the loss of sensation. The pains are due, no doubt, to slight changes in the posterior root fibres or peripheral nerves.

The loss of knee-jerks is caused by lesion of the reflex arc in its sensory portion—in the intra-medullary fibres of the posterior root, or their reflex collaterals, or the peripheral sensory nerves. According to Sir William Gowers, affection of the afferent muscular nerves is the cause of the loss of knee-jerks and hypotonus, and is one cause of the ataxia.

Co-ordination of movement in health is chiefly an acquired function; it is learned through practice. The movements of infants are at first ataxic and only by practice is co-ordination acquired; by practice the child learns to walk. Centripetal impulses come not only from the skin, but also from the deeper parts, muscles, joints, etc., and the movements are further regulated by vision. The disturbance of co-ordination in tabes is caused by degeneration of centripetal fibres which regulate the movements. It does not follow that ataxia is due to changes simply in the sensory nerves of the skin, since with complete anæsthesia of the skin there may be no ataxia and in some cases of tabes no affection of cutaneous sensation can be detected by the most delicate tests. Marked ataxia has been observed in disease of the posterior nerve roots, as in cases of tumours of the posterior roots, recorded by Dr. Hughes Bennett.

According to Frenkel when ataxia is present in tabes the sensation of passive movements at the joints of the limbs (joint sensation) is always affected. But Ferrier does not think the two conditions are so closely related as Frenkel believes.

Tabetic ataxia appears to depend on changes in the intra-medullary fibres of the posterior roots and changes in the afferent muscle nerves, which cause impairment or loss of centripetal impressions, especially those from the deeper structure, muscles, tendons and joints. The hypotonus of the muscles also aids in producing ataxia, since the resistance offered by the opponents of the contracting muscles is less than normal, and hence the impressions of muscle sense from the limbs are defective.

(The causation of ataxia is carefully discussed by Ferrier in his Lumleian lecture, 1906.)

The trophic changes in the bone, skin, and joints are probably the result of degeneration of peripheral nerves in these structures. Changes

in the neighbourhood of the vagus nucleus may be the cause of gastric and laryngeal crisis.

The pupils in general paralysis and tabes are often irregular in outline and not infrequently are eccentrically situated. This condition often precedes the Argyll-Robertson pupil. These irregularities (adhesions and congenital anomalies being excluded) are of diagnostic value and are very suggestive of tabes, general paralysis or syphilis.

By irritation of the short and long ciliary nerves in dogs, Piltz produced similar irregularities in the pupils.

It appears probable that the alteration in the shape of the pupils in tabes is caused by degeneration of the ciliary ganglion and short ciliary nerves.

The seat of the lesion causing the Argyll-Robertson pupil has been disputed. It is not in the nucleus of the third nerve. It has been attributed to lesion of fibres connecting the corpora quadrigemina with the nucleus of the third nerve: others believe the lesion to be in the cervical region of the cord; but the changes found by Marina in the ciliary ganglion are important and may be the cause of the loss of the light reflex.

Marina comes to two conclusions as the results of his pathological observations—(1) In all cases of tabes and general paralysis of the insane with affection of the pupillary reflex, the ciliary ganglion (and usually its nerves also) present pathological changes; but these structures are normal when the light reflex is present. Chronic changes leading to complete chromatolysis were found in the nerve cells of the ganglion. (2) In all cases with immobility of the pupils the nuclei of the third nerves were normal.

Ferrier thinks that there is "nothing improbable in the assumption that in tabes the ciliary ganglion and ciliary nerves are so affected that, though they cannot transmit the reflex impulse of light to the sphincter pupillæ, they can readily allow the more powerful stimulus associated with accommodation to pass through."

(The subject is discussed carefully by Dr. Ferrier in the Lumleian lectures at the Royal College of Physicians London, 1906.)

Treatment of Tabes.

Medical treatment is often of service, though there is no drug which can be regarded as a cure for the disease. In the first place it should be borne in mind, that tabes is occasionally simulated by spinal syphilis (pseudo-tabes); and in the latter affection, there is a possibility of cure by suitable anti-syphilitic treatment.

As regards true tabes, it is important to remember that in many cases the affection ceases to progress: and in the progressive cases the advance of the disease is usually very slow, and death generally occurs from some complication. Much can be done to prevent these complications and to relieve troublesome symptoms. Further, for one

prominent symptom—ataxia—we have now a method of treatment which often gives excellent results.

The question of the **prevention** of the diseases is only likely to be considered in the case of persons who have suffered from syphilis and dread the development of tabes. Probably a thorough treatment of syphilis at its early stage diminishes the risk of tabes occurring at a later date. According to Prof. Erb the most important part of the prophylactic treatment of tabes consists in "the timely, thorough and sufficiently long treatment of syphilis at its onset." But on this point there is considerable difference of opinion, and the statistics of a number of observers appear to show that early anti-syphilitic treatment does not always prevent tabes.

Whilst syphilis is the most important factor in the causation of tabes, it is believed by many that there are contributory causes, which favour the onset of the disease in those who have suffered from syphilis. Hence it is important for such persons to endeavour to avoid these injurious influences, i.e. great exposure to wet and cold, physical over-exertion, excessive mental work, sexual excess, and excess with respect to alcohol and tobacco.

When tabes has developed it is important, in the first place, to decide whether **anti-syphilitic treatment** should be given, if there be a history of previous syphilis. In all cases in which there is the least doubt in the diagnosis between early tabes and pseudo-tabes due to spinal syphilis, anti-syphilitic treatment should be given; but if the diagnosis of tabes is certain there is much difference of opinion as to the value of anti-syphilitic treatment. Many writers state that they have found it useless, but others have obtained improvement in certain cases. Professor Erb, who has devoted much attention to the subject, believes that anti-syphilitic treatment is of value in many cases; and that it is usually unattended with bad effects; but he admits that in a not inconsiderable number of cases it does not produce any good results. Erb advises it :—

1. In cases in which tabes has developed within a comparatively short time of the syphilitic infection.[1]

2. In cases in which syphilitic symptoms or lesions are still present, or in which symptoms of cerebral or meningeal syphilis occur.

3. In cases in which the patient has not previously had a satisfactory treatment for the syphilis.

There can be no doubt that in tabes anti-syphilitic treatment has not the effect which it has in tertiary syphilis; in well advanced cases of tabes it is usually of no service: and many writers believe that it is sometimes injurious. According to Erb anti-syphilitic treatment is contra-indicated in advanced cases of tabes, in dyspeptic patients, in those who have already had repeated courses of antisyphilitic treat-

[1] Collins recommends antisyphilitic treatment when tabes has developed within five years of syphilitic infection.

ment without effect, and in patients who show intolerance of mercury and potassium iodide.

The form of mercurial treatment recommended by Erb is the inunction course. One inunction may be given daily for one or two months. Then the course should be discontinued. Erb advises several inunction courses with intervals of six to twelve months. During the intervals he gives tonics or potassium iodide. He does not consider the latter drug so valuable as mercury, but thinks it is specially indicated in tabes when symptoms of tertiary syphilis are present, when there are severe pains, and when the symptoms are progressing rapidly. The dose should be 20 to 60 grains in the twenty-four hours : it may be given in milk or alkaline water, or with aromatic spirits of ammonia. Sodium iodide is preferred by many in place of potassium iodide.

When optic atrophy is present mercury and iodides should not be employed.

General Treatment.—It is important for the patient to live a regular life. Excesses of all kinds—sexual excess, bodily over-exertion, mountain climbing, great exposure to wet and cold, excessive mental work and excess with respect to alcohol and tobacco—should be avoided. If the patient be in the pre-ataxic stage it is usually advisable that he should not marry. Cold and wet increase the pains and inco-ordination, hence a warm dry climate is best.

Rest.—There can be no doubt of the great value of rest in bed for several weeks in early cases which are rapidly progressing, in cases in which there has been much bodily over-exertion or sexual excess, in cases in which ataxia has developed rapidly, and in cases in which there is great weakness or increase in the frequency of pains. Many authors have drawn attention to this point. The writer has seen good results from the rest treatment,and believes that this has been the explanation of the striking temporary improvement which has occasionally followed, and been attributed to, the use of simple bitter tonics, etc., which cannot be regarded as having any influence on the disease.

The patient should attend to the condition of the bladder and bowels. The urine should be passed at least four times in the twenty-four hours, and the bowels should be kept regular.

Drug Treatment.—Sir William Gowers and Dr. James Taylor think that arsenic—(3 to 7 m. of liquor arsenicalis)—and aluminium chloride (2 to 4 grains) twice or three times a day, are the drugs which most frequently do good.

Many writers of experience think that nitrate of silver is sometimes of service, and others believe that strychnine and nux vomica sometimes act favourably.

Potassium or sodium iodide is worthy of trial when shooting pains are troublesome.

Nitro-glycerine is recommended by Osler in cases of tabes in which the arterial tension is increased.

In Germany treatment by *baths* at Oeynhausen and Nauheim is thought to be of some service by several careful observers. Hot, cold, and vapour baths should be avoided ; but tepid and warm baths are suitable, and are especially recommended by Starr for two or three months twice or three times a year.

Counter-irritation of the spine (by the thermo-cautery) is believed by Gowers to be sometimes useful, especially when there is a tendency for the symptoms to increase, or when there is spinal pain or tenderness. It has been strongly recommended by McLane Hamilton.

Massage is recommended by Starr at an early stage and at the last stage. Kouindjy has carefully studied the value of massage in tabes. He considers that massage ought to be employed with extension, in cases of tabes without ataxia ; and with extension and re-education of the muscular movements in cases of tabes with ataxia. In cases of tabes without ataxia, massage treatment ought to precede extension. In cases of tabes with ataxia, massage ought to follow the re-education of movements. Frenkel, however, has recently written decidedly against massage.

In many tabetic patients there is wasting and general impairment of health. Hence it is very desirable to keep up the general nutrition by favourable hygienic conditions, and by abundant diet, or over-feeding. Cream, cod-liver oil, and fatty articles of food may be given for this purpose. In addition the open-air treatment has been specially recommended by Determann.

The **Suspension treatment** for tabes was first employed by a Russian physician, Motschutkowski. The patient is suspended by means of an apparatus like that employed during the application of a Sayre's plaster of Paris jacket. The duration of the suspension should be half a minute increased up to five minutes. This treatment has been extensively employed, but in England it is now very rarely used. Many careful observers have obtained no good results and a considerable number of serious accidents have occurred during the treatment, some of which have been fatal. Nevertheless, there is some slight evidence in its favour. Motschutkowski still recommends it. He has employed it in 993 cases of tabes, 207 of which have been improved thereby ; the improvement being noted in the gait, the muscular tone, the general condition, the muscular sense, paræsthesia, neuralgic pains, sexual power and bladder functions. Motschutkowski recommends that the patient should be suspended at least 100 times, and that the suspension should be made three or four times a week. He has never seen any bad results. Erb recommends it in emaciated individuals, in cases with shooting pains, gastric crises, bladder and sexual weakness, girdle and cuirass sensations, and commencing ataxia. Charcot thought it diminished the shooting pains and the impotence. Oppenheim also has occasionally obtained good results. In v. Leyden's clinic an apparatus devised by P. Jacob is employed, in order to

prevent the risk of accidents during the treatment. This apparatus consists of an inclined plane on which the patient is placed and suspended, the head being fixed by a band passing under the chin. The angle at which the plane is inclined is at first small ; afterwards it is made greater and the traction on the spine caused by the weight of the body is thus increased. By this method of treatment v. Leyden found that the shooting pains and gastric crises were diminished. The suspension treatment should not be employed when arterio-sclerosis is present, nor when the disease is complicated with bulbar symptoms or general paralysis of the insane.

Gilles de la Tourette and A. Chipault have introduced a method in which the spinal cord is supposed to be stretched, by flexion forwards of the spine. The patient being seated on a table with the legs extended and adducted, the head is drawn forwards forcibly towards the knees. The patient remains in this position for about ten minutes. This treatment is carried out twice a day.

Stretching of the sciatic nerve was formerly frequently practised, but experience has shown it to be useless.

The value of **electrical treatment** is probably slight, but some writers think that galvanism to the spine is of service. One large electrode should be placed on the neck and the other at the lumbar region. The strength of the current should be 5–8 milli-ampères. The electrodes remain fixed, or the upper one can be drawn down the vertebral column. The duration of each application should be about five minutes, and the treatment should be given daily, or three times a week for many months (Oppenheim). An ascending current—stable or labile is recommended by F. Moritz.

Oppenheim thinks that galvanism should be tried in every case ; he has found that it often produces a decided improvement in the subjective symptoms, and that sometimes there are also objective signs of improvement. F. Moritz states that occasionally the pain, the ataxia and bladder symptoms diminish under this treatment.

Treatment of Complications.—For the relief of attacks of severe *shooting pains* rest in bed is advisable, and local applications are worthy of trial—such as fomentations, rubbing the painful region with chloroform, or the application of lint which has been soaked in chloroform. Occasionally a warm bath, Franklinic electricity, and the faradic brush are of service in diminishing the pains. Internally several drugs have been found useful, especially antipyrin, antifebrin and pyramidon ; also phenacetin, exalgin, salipyrin, aspirin, sodium salicylate, and aluminium chloride have sometimes been of service. Potassium iodide is useful in cases in which severe pains are frequent. The most certain remedy is the hypodermic injection of morphia, but this should only be used as a last resort, since there is a great danger of the morphia habit being formed. The patient should never be allowed to use the morphia syringe himself.

Sir William Gowers recommends the hypodermic injection of cocaine ($\frac{1}{12}$ gr. increasing up to $\frac{1}{3}$ gr.) for pains that are superficial in character. Dejérine recommends a powder consisting of $7\frac{1}{2}$ grains of antipyrin and $4\frac{1}{2}$ grains of phenacetin. Santonin has been found useful by Negro and others. Oppenheim recommends pyramidon and aspirin as two of the best drugs for the relief of the pains of tabes. Schultze regards pyramidon as the best drug for the relief of the pains; Sir William Gowers thinks antifebrin (acetanilide) is the most useful; he believes that aluminium chloride lessens the tendency for the occurrence of pain, and that aspirin and sodium salicylate have a similar action.

Antipyrin (in 10 gr. doses) is the drug which I have used most frequently, and it has often been of distinct service.

The positive pole of a galvanic battery saturated with a solution of cocaine, 6 to 10 per cent., abolishes the sensibility of the skin to pain, and may be of service.

During the *gastric crisis* complete rest is necessary. A little ice may be sucked, but if any food can be taken the amount should be very small. Feeding entirely per rectum is sometimes necessary. Morphia given by rectum, and morphia injections during the attacks, are sometimes of service and may diminish the revulsion to food. Chloral may be of service by giving sleep and rest for many hours. Erb has often found benefit from the application of the galvanic current to the epigastrium. Others have obtained relief by a blister, by ice, or by methylchloride spray applied over the epigastrium. Internally cocaine hydrochloride, chloroform water, extract of Indian hemp, cerium oxalate, codeia, dionin, and belladonna have also been employed—sometimes apparently with advantage. Between attacks the patient should be fed up as much as possible, so that he may be able to resist the great loss of strength produced by the crises (v. Leyden).

In the *laryngeal crises* the inhalation of chloroform or ether, painting the larynx with cocaine, morphia injections, nitrite of amyl and nitroglycerine have been found to be of service.

For *weakness of the bladder* wall and of the sphincters, strychnia has been found useful. Electricity (galvanism or faradism) to the perineum, is of service in some cases. It is also advisable to aid the emptying of the bladder by pressing with the hand on the lower part of the abdomen at the end of micturition. The use of the catheter is desirable when the bladder cannot be completely emptied: and when cystitis has developed, it may be necessary to wash out the bladder every day or every second day. For this purpose boracic acid lotion or chloroform water (as recommend by v. Leyden) may be used. Also when cystitis has developed urotropin should be given internally.

In cases of tabes in which *optic atrophy* is present I believe it is important not to give iodide of potassium or mercury. From my own experience I believe these drugs increase the rapidity of visual failure. Many physicians have come to the same conclusion. We have no treat-

ment which can cure or even arrest the optic atrophy and visual failure but a thorough trial of hypodermic injections of strychnia should be made, since several observers believe that it has a slight beneficial effect. (Sir Wm. Gowers recommends the injection of nitrate of strychnia daily for six months—dose $\frac{1}{30}$ to $\frac{1}{15}$ of a grain; afterwards for one week in five.) It is also desirable that eye-strain in the patient's work should be carefully avoided in all cases of tabes, and that tobacco smoking should be restricted.

The condition of the feet may require careful attention. Corns should not be cut, as a perforating ulcer may be produced by a corn being cut too deeply. The epidermis should be softened by an alkali and the surface rubbed away with pumice stone.

When a *perforating ulcer* has developed, rest in bed or on the sofa, and the application of a paste of iodide of starch may be recommended.

Chipault and others have recorded cases in which stretching of the posterior tibial or plantar nerves has caused old standing perforating ulcers to heal quickly.

J. Crocq has employed faradism with good results. A small electrode is placed on the posterior tibial nerve behind the internal malleolus, and a larger one on the sole of the foot, just behind the ulceration. A strong current is passed for a quarter of an hour through the parts.

Charcot's Joint Disease.—Rest is of service at first: afterwards a special mechanical apparatus is sometimes useful. Puncture of the joint is sometimes of service. The best results are obtained when ankylosis occurs in a good position. Hence arthrodesis or excision may be desirable for obtaining the ankylosis. In some cases when the limb is quite useless amputation is desirable.

Deformities at the knee and other joints can sometimes be corrected by a suitable splint apparatus or support.

The tabid club-foot (pied bot tabétique of Joffroy) may be prevented from developing by using a cradle to avoid the pressure of the bed clothes on the dorsum of the foot.

Treatment for Ataxia.—The greatest advance in the palliative treatment of tabes is the method of re-education of the movements of the ataxic limbs, by a long course of compensatory exercises carefully carried out. It was first introduced by H. S. Frenkel in Switzerland: and it has been also largely employed by Goldscheider in Berlin and by many others. There can now be no doubt that the ataxia of tabes is often greatly diminished by this system of careful training. It is, of course, chiefly of value in the second or ataxic stage of the disease; but even in the third or "paraplegic" stage it may be of some service. The method requires, however, great patience and attention on the part of the medical man who supervises it, and perseverance and intelligence on the part of the patient.

It is important to remember that the object of the treatment is *not*

to increase the strength of the muscles by gymnastic exercises and muscular movements. The aim of Frenkel's treatment is to *re-educate* the movements of the ataxic patient. In other words by repeated practice the patient learns again to co-ordinate his movements—to co-ordinate the action of his muscles.

The infant is ataxic in most of his movements : the power of co-ordinating the movements of the limbs, and the movements necessary in standing erect and walking, must be acquired in infancy and childhood. In walking a very marvellous and complicated co-ordination of movements must be developed. Not only the action of the leg muscles, but also those of the trunk, must be very accurately co-ordinated.

Frenkel's treatment can directly influence only one symptom of the disease—ataxia. By repeated practice, in this method of treatment, the ataxic patient learns to perform definite movements with care and precision, beginning with the simple movements and gradually proceeding to those which are most difficult. By concentration of the attention on the movements, by making the best use of the sensation which still remains, and especially by the aid of vision, the patient learns to co-ordinate again the movements of the ataxic limbs—just as a normal individual may learn, by practice, to skate, to cycle, or even to walk upon a tight rope, by the education and fine co-ordination of his muscular movements.

In carrying out Frenkel's re-education treatment it is very important that over-exertion should be avoided. The condition of the pulse should be watched. Usually the pulse frequency is increased, even when the movements performed are only of two or three minutes' duration. Before a new movement is undertaken it is advisable to wait until the pulse beats diminish to the normal number. In tabes the feeling of tiredness, after long exercise, is often lost or diminished ; hence the patient's feelings are no indication of the time which can be devoted to the exercises without causing over-strain. Frenkel recommends that the exercises should be carried out twice a day only, and that they should not be continued for more than 5-15 minutes. This time includes the periods of rest.

The treatment should be continued for several months. Improvement is usually obtained, but the extent thereof varies, and depends on the duration of the treatment more than on the severity of the ataxia.

When the ataxia has developed rapidly, a period of rest is first advisable, and the Frenkel's treatment is best postponed for a short time.

Unfavourable complications preventing good results are : attacks of pain lasting for days, frequent gastric and intestinal crises, marked acceleration of the pulse, a high degree of hypotonus with stretching of the joint ligaments, joint affections and luxations. Joint deformities due to hypotonus of the muscles may require correction before the exercises can be carried out. Blindness, from optic atrophy, was formerly

regarded by Frenkel as a complication rendering the exercise treatment hopeless, but recently with a modified method he has obtained some improvement.

The exercise treatment has rendered the prognosis in tabes much more favourable ; many complications which often occur through deficiency of exercise can now be avoided.

Massage—which is often advised in conjunction with the exercise treatment—has now been abandoned by Frenkel, since he has not found it of value.

It is always desirable that some one should closely follow the ataxic patient during the walking exercises, in order that accidents and falls may be prevented. Tabetic patients sometimes fall down very suddenly, through a sudden "giving way" of the knees, and not infrequently fractures are thus produced. In severe cases of ataxia the patient may require to be supported by means of a girdle belt fixed round the chest, during the exercises.

The room in which the exercises are carried out should be a long one (Frenkel recommends one about 25 yards long). Black or coloured stripes should be painted on the floor, or better still on linoleum, which can be taken up when no longer required. Frenkel recommends the following : (1) One stripe should be about 9 inches wide. (2) Another about half that width. Two similar stripes (3 and 4) divided by cross lines, 27 inches apart. (This is the length of a full step.) Each section is further equally sub-divided into half and quarter steps. (5) A zigzag stripe 9½ inches wide, the length of the segments being 27 inches. (6) Two other stripes on which pairs of footprints are painted. Other more complex footprints and floor markings are also used. Special apparatus for exercises of the legs in bed and for exercises of the arms are shown in Fig. 151.

For the legs four forms of exercises are employed :—

1. Those practised in the recumbent position (in bed).
2. Those practised when the patient is in the sitting posture.
3. Those performed whilst the patient is standing.
4. Those carried out during walking.

1. The first group of movements, performed in bed, consists of flexion and extension of the knee, abduction and adduction of the legs, touching the knee of one leg with the heel of the other, drawing the heel of one leg along the shin bone of the other, etc., etc.

Three forms of apparatus are also employed by Frenkel for certain movements of the legs (Fig. 151)—a cross-bar, a board with eight holes, and vertical boards with heel rests. The heels are placed upon the cross-bar, into the holes, and on to the rests in the various exercises. Three other forms of apparatus which have been used are shown in Fig. 151.

2. The exercises performed in the sitting posture are :—raising the thigh, with the knee flexed, and putting the foot on the ground firmly ;

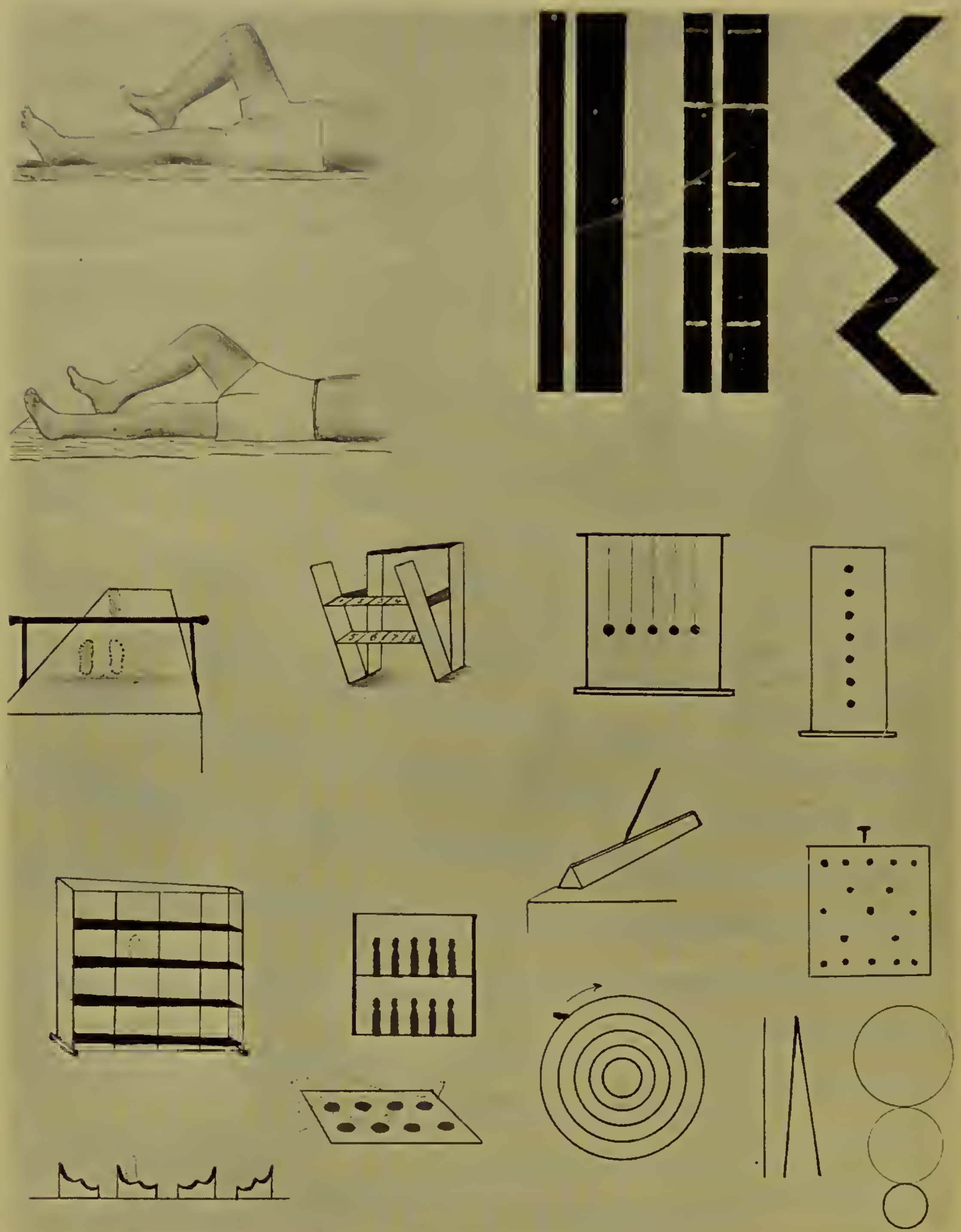

FIG. 151.—Diagram to illustrate the Exercise Treatment for Tabes Dorsalis.

In the left half of the illustration.—At the upper angle the first figure shows the movement of touching the knee with the heel of the opposite leg; the second figure, the drawing of the heel of one leg along the tibia of the other. Below the last figure is a diagram of the bed with a cross bar (deep black). The foot is placed on and under this cross bar. To the right of this figure is an apparatus like an inverted small step ladder: on the cross pieces of the apparatus are squares marked 1 to 4, and 5 to 8; the heel is placed on these squares in succession. Below the two figures last described are:—An apparatus with sixteen square compartments, into each of which the foot is placed in succession; and another apparatus with two rows of five skittles, which are knocked over in succession by the foot. At the lower left angle are four wood vertical blocks, each having two grooves into which the heels are placed. To the right of the last figure is a board with eight holes, into which the heels are placed in succession.

In the right half of the illustration.—At the upper part are a narrow and a broad black stripe on which the patient places the feet in walking, similar stripes with divisions and a zigzag stripe. Below these figures is an apparatus with five balls attached by strings to a cross bar. In the treatment of ataxia of the arms the patient practises catching these balls when they are swinging. To the right of the last figure is the diagram of a board with round holes, into which the patient places the tip of the index finger. Below the last two figures are:—A prism with a groove at its upper margin in which the patient slowly draws the point of a pencil; and to the right of this figure is represented a board with holes, into which the patient inserts a small plug. At the lower right angle of the illustration are circles and lines drawn on paper, which the patient copies. Just to the left of the figures last mentioned is a small circle surrounded by four others. The patient moves the tip of the index finger along these circular lines.

putting the foot on to footprints on the floor; getting up and sitting down, etc.

3 and 4. Of the standing and walking exercises, which should be performed very slowly and with great attention, the following may be mentioned :—standing on footprints and on floor markings; walking along the broad stripe (Fig. 151) with short steps and then with long steps; walking sideways and walking backwards; turning round; walking on the zigzag stripe; walking on the narrow stripe; walking on cross divisions of the stripes and on footprints, etc.

For the exercises of the arms the following forms of apparatus are employed : (1) a prismatic block, with grooved upper margin in which the patient draws a pencil (*see* Fig. 151); (2) a perforated board into the holes of which the tip of the index finger is placed; (3) a board with holes and pegs (the patient placing the pegs into successive holes); (4) swinging balls which are to be caught whilst in movement; (5) round discs, like coins, which the patient piles up one upon the other; (6) simple diagrams on paper, which are to be copied.

The results of the exercise treatment vary considerably; but in many cases they are satisfactory. The improvement has also a most important mental effect, it gives to the patient new hope and courage.

In a case under my care, a few years ago, the results were exceptionally favourable. The symptoms of tabes were typical, and for five years the patient had been confined to bed on account of the ataxia. When he came under treatment he was quite unable to walk even with assistance, and was unable to stand, even when supported well on each side, owing to ataxia. For eleven weeks the treatment was carried out. First he performed simple movements of the legs in bed with precision. These were increased in number, and in time were made more complicated. Then, afterwards, walking movements were carried out with assistance on either side, afterwards walking with the aid of a stick, and finally walking alone. At the end of the treatment the ataxia had not disappeared, but it had diminished so much that he was able to walk about the room alone, and for short distances without the aid of a stick. Since that time, five years have elapsed and the improvement has been maintained.

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FRIEDREICH'S DISEASE—HEREDITARY ATAXIA

This affection was first described by Prof. Friedreich, of Heidelberg.

Etiology.—The disease has a well-marked tendency to affect more than one member of the same family ; two or more children of the same parents may suffer from it, whilst the other children in the family are unaffected. Much less frequently the disease appears in successive generations. Generally the parents are not affected. Occasionally isolated cases are met with. The age of onset is most frequently about the 7th or 8th year ; sometimes later, 12th–16th years ; but seldom after this age, and never after 25. In the same family, the onset of the disease is usually about the same age, in the case of each member affected (Soca). Infectious diseases may act as exciting causes.

Symptoms.—The first symptom which attracts attention is usually unsteadiness in walking. The ataxia is slight at first, but in course of time it gradually increases. The gait becomes slow and uncertain ; the patient looks at his feet when walking ; the steps are rather short, the feet are widely separated ; the patient deviates now to the right, now to the left, and the gait somewhat resembles that of a drunken man. The ataxia is more like that of cerebellar disease than of tabes. There is not the rapidity and excess of movement which is common in tabes,

also the oscillations of the body in walking are greater than in tabes. At a later date there is ataxia on standing (static ataxia); the patient stands with the feet widely apart; he is unsteady; and the body sways from side to side. The head and upper part of the body are bent forwards.

In some cases the unsteadiness is increased when the eyes are closed; in others closing the eyes has little influence on the unsteadiness.

Ataxia of the arms develops at a later date than that of the legs. There is inco-ordination of the movements of the arms and fingers; there is difficulty in using a spoon, in writing, in buttoning the clothes, and in picking up small objects.

Often there are also irregular nodding movements of the head and neck, somewhat like those seen in disseminated sclerosis.

At first the muscular power is unimpaired, and there is no atrophy of muscles.

The speech becomes affected, but this symptom is usually much later in its development than the ataxia. The speech is slow, thick and indistinct, at times hesitating; syllables are elided, and some words are pronounced more quickly than others. When the tongue is protruded tremulous or jerky movements are sometimes observed. There are, in some cases, irregular, restless, or choreiform movements of the arms, but they are not so constant and violent as in true chorea. Occasionally athetoid movements have been observed.

Nystagmus is present in many cases, but not in all; it occurs chiefly in lateral or upward movements of the eyeballs. The pupils generally react normally. Optic atrophy does not occur (or it is exceedingly rare), and paralysis of ocular muscles is very seldom seen.

The knee-jerks usually disappear at an early period; in rare cases the knee-jerks have been present. Ankle-clonus is absent.

The superficial reflexes are usually present; but sometimes the cremasteric reflexes are absent. The plantar reflex is of the extensor type (Babinski's reflex), a sign of degeneration of the crossed pyramidal tract. Muscular hypotonus, like that of tabes, is often present. The bladder and rectum are not generally affected.

Fig. 152. — Foot; Friedreich's Disease. Note hyper-extension of toes, especially big toe; prominence of tendon of flexor hallucis longus; swelling on dorsum of foot; hollow sole of foot.

Lateral curvature of the spine is common, sometimes there is kyphosis, or kyphoscoliosis.

The feet very frequently present a well-marked deformity. The whole foot is dropped and is in the position of talipes equinus or equinovarus; the foot is shorter than in health in the antero-posterior direction; the sole is hollowed out—pes cavus; the dorsal surface of the foot is

very prominent; and the toes are hyper-extended. The hyper-extension of the big toe is especially marked, the toe being extended at the metatarso-phalangeal joint, but flexed at the phalangeal joint. The tendon of the extensor proprius pollicis is very prominent. In some families, in which several members have successively suffered from the disease, this prominence of the tendon of the extensor proprius pollicis has been the first sign of the affection which has attracted the attention of the parents.

In many cases, even at a late period, sensation is normal; and when sensory symptoms are present, they are usually slight.

Shooting or severe pains are exceedingly rare; but sometimes there are slight dull or aching pains in the limbs; occasionally there is numbness of the hands and feet, or slight anæsthesia in the legs.

There are no trophic changes in the skin or joints.

As already mentioned, there is no paralysis at the early stage; but at a late period there is often rigidity with some loss of power in the limbs, most marked in the legs. At a late period slight wasting of muscles may occur. In rare cases there has been wasting of the small muscles of the hand, causing the "claw-shaped hand."

The electrical excitability is only slightly diminished; the muscles react to faradism and galvanism, and there is no qualitative change in the galvanic reactions.

The mental condition is not affected, or there is simply slight mental impairment at a later stage.

Visceral crises are absent. Newton Pitt and others have pointed out that symptoms of cardiac failure (muscle failure) have often been observed.

The course of the disease is very chronic. Recovery does not occur; but often the patient lives for many years, frequently for 30 years after the onset of the disease. In many cases the disease has not apparently shortened life; but death may occur at the end of 10 to 12 years, generally from some intercurrent disease.

Diagnosis.—From tabes dorsalis the diagnosis is usually easy. The points of difference are shown in the following table:—

Friedreich's Disease.	Tabes Dorsalis.
Onset in childhood.	Usually after the age of twenty years.
Often several children in the same family affected.	Usually no family tendency.
History of syphilis, congenital or acquired, very rare.	Frequently history of syphilis.
Absence of lancinating pains.	Shooting pains are usually present and precede ataxia for several years.
No pre-ataxic stage.	Long pre-ataxic stage.
Sensation normal, (or only slightly affected in rare cases).	Sensation frequently affected.
Nystagmus and oscillatory movements of head and neck common.	Almost always absent.
Pupils usually react normally.	Argyll-Robertson pupil present in most cases.
Speech affected (*see* description).	Usually not affected, or only slightly affected.

Sensory cranial nerves not affected. Optic atrophy absent.	Optic atrophy occurs in 10–15 per cent. of cases. Other cranial nerves may be affected.
Scoliosis and peculiar deformity of the foot very common.	Absent.
"Crises" and trophic changes absent.	Often present.
Extensor form of plantar-reflex (Babinski's reflex) present.	Plantar reflex of normal flexor type.

The difficulty of diagnosis occurs chiefly in the rare cases of tabes occurring in early life (juvenile tabes), but the differences in the symptomatology already indicated are usually sufficient to separate the two affections. Also evidences of syphilitic disease of the teeth, interstitial keratitis, and reflex iridoplegia would be in favour of tabes.

The points of difference from disseminated sclerosis are indicated in the following table :—

Freidreich's Disease.	Disseminated Sclerosis.
Age of onset, 7 to 16.	18 to 40.
Often several members of the family affected.	Usually no family tendency to the disease.
Scoliosis and peculiar deformity of the foot common.	Absent.
Optic atrophy and visual defects absent.	Frequently present (often partial).
Knee-jerks usually absent.	Rarely absent, usually increased ; often ankle-clonus present.
Progressive ataxia of limbs.	Frequently tremor of arms on voluntary movement.
Gait ataxic.	Gait often spastic ; but if ataxic at onset usually knee-jerks present ; and limbs often rigid.
Very chronic course.	More rapid course.

Cerebellar tumour often causes an ataxic gait ; but headache, vomiting, and double optic neuritis are common symptoms, whilst they are absent in Friedreich's disease ; also the onset of the symptoms is much more gradual, and the duration of the affection much more prolonged in Friedreich's disease. Ataxia of the arms is common in Friedreich's disease, rare in cerebellar tumour.

Ataxic paraplegia and other forms of combined postero-lateral sclerosis are usually easily separated from Friedreich's disease by the presence in the former disease of increased knee-jerks, ankle-clonus, and rigidity of the legs, and by the absence of nystagmus and speech affections. But occasionally the knee-jerks are present in Friedreich's disease, and may be lost in the other forms of postero-lateral sclerosis. In such case the early age of onset, the family tendency are important points in favour of Friedreich's disease.

Pathological Anatomy.—To the naked eye the spinal cord appears smaller and thinner.

Microscopically sclerosis is found in the posterior and lateral columns of the cord. The changes are symmetrical and are most marked in the dorsal region. The posterior columns are sclerosed at all parts of the cord, and the changes in the column of Goll are more intense than those in Burdach's column. The sclerosis in the posterior columns is sometimes

less marked in the lumbar region than at a higher level, but the lumbar posterior root zones are affected. In the cervical region both the columns of Goll and Burdach may be affected. Generally the posterior root zone is markedly affected; and it does not escape in the cervical region as in many cases of tabes. The endogenous tracts in the spinal cord are less affected than those containing fibres from the posterior roots.

In advanced cases all parts of the posterior columns may be affected except the posterior ventral field (a narrow strip of white matter close to the grey commissure and neck of the posterior horns) and the centrum ovale of Flechsig; also the postero-external field of the posterior columns is less affected than other parts which contain exogenous fibres.

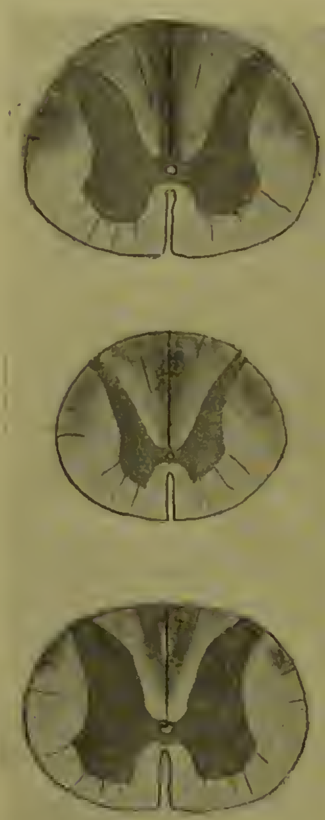

FIG. 153.—Spinal Cord; Friedreich's Disease. Deeply shaded parts of white matter are degenerated. Uppermost figure = cervical region; middle figure = dorsal; lowest = lumbar (diagrammatic).

Degeneration is found in the region of the crossed pyramidal tracts. The sclerosis is not limited to the crossed pyramidal tracts, but also involves the direct cerebellar tracts throughout their entire length, and in advanced cases the tracts of Gowers may be affected. Moreover the degeneration extends forwards on the surface of the cord and sometimes there is distinct degeneration in the anterior pyramidal tracts (*see* Fig. 154).

In the columns of Clarke (posterior vesicular) there is always degeneration or atrophy of nerve cells and partial degeneration of nerve fibres. These changes are associated with the sclerosis in the direct cerebellar tracts. The zone of Lissauer has been normal in some cases, in others it has been affected. The grey matter and the nerve cells of the anterior horns have generally been normal.

The posterior nerve roots may present partial or marked degeneration of their fibres; but they have been reported normal in some cases. The anterior roots are normal. In some cases the pia mater has been found thickened, especially over the region of the posterior columns; in other cases it has not been affected. Degeneration of the cells of the posterior root ganglia has been observed.

In many cases on record the peripheral nerves have not been examined; in some cases they have been normal, in others degenerated.

In the sclerosed tracts of the spinal cord there is an increase of the neuroglia (which is converted into dense fibrillar tissue) along with atrophy of both the medullary sheath and axis cylinder of the nerve fibres. Dejerine, Letulle and others have drawn attention to the structure of the

sclerosed tissue. In cases which they have described the sclerosed neuroglia tissue has consisted of fibrillæ intercrossed in different directions and forming "whorls." But this condition is not always present.

The degeneration is most intense in the posterior columns and direct cerebellar tracts.

In one case Friedreich reported atrophy of the cells of the posterior pyramidal nucleus and some degeneration of the restiform bodies.

Changes have been found in some cases in the motor tracts in the lower part of the medulla.

Atrophy of the cerebellum was found in a case reported by Senator, and this condition has been recorded in several cases presenting symptoms somewhat allied to Friedreich's disease; but in most cases recorded the cerebellum has been found normal (*see* Mackay's collected records).

In ataxic paraplegia there are similar changes in the posterior columns; but the posterior root zone in the lumbar region is free, or is not affected so much as in Friedreich's disease. Also the sclerosis of the posterior columns is not so marked in the lumbar region in ataxic paraplegia as in Friedreich's disease. Further the posterior roots are not affected in ataxic paraplegia, but are often degenerated slightly or markedly in Friedreich's disease.

FIG. 153A.—Diagram showing Distribution of Anterior and Posterior Arterial System of the Spinal Cord. Area of white matter supplied by posterior arteries is shaded. P = posterior intermediate septal artery.

The early age of onset and the family tendency of the disease suggest that the cause is a congenital defect. Sir W. Gowers thinks the disease is due to a congenital morbid tendency of development, by which the affected nerve elements have a briefer period of vital endurance than the other tissues of the organism (abiotrophy).

It is worthy of note that the degeneration is most intense in that part of the transverse section of the cord, which is supplied by the peripheral arteries of the cord, whilst the part suppled by the central arteries is not affected. The former is the less vascular region, and this fact may account for the localisation of the changes. Nerve fibres having an impaired power of resistance may degenerate first in the part of the cord least freely supplied by blood (*see* Fig. 153A).

Dr. Newton Pitt has drawn attention to vascular change in Friedreich's disease; he thinks that the cord changes are probably associated with "an inherited tendency to general early vascular deterioration."

Treatment.—The disease is incurable and the treatment can only be symptomatic. At an early period Frenkel's exercise treatment may be of some service in diminishing the ataxia. Strychnine and tonics are often given, but usually without benefit.

Hereditary Cerebellar Ataxy.

This is the name given to a group of cases described by Marie which somewhat resemble Friedreich's disease clinically. Marie regards the symptoms as the result of a congenital atrophy of the cerebellum. More than one member of the same family may be affected; though isolated cases are met with.

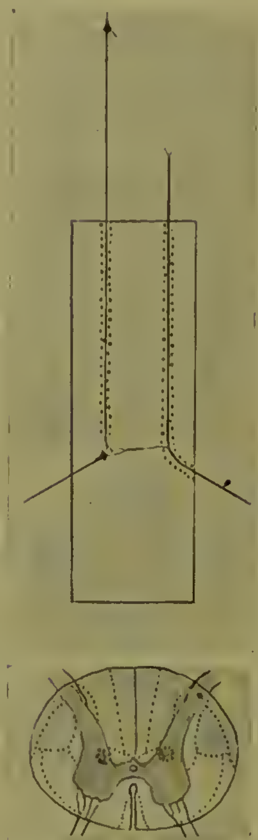

Fig. 154.—Friedreich's Disease. Lower figure = transverse section of cord; degenerated parts are shaded. Upper figure = longitudinal section of cord. To the left a fibre of the anterior nerve root and its cell of origin (lower motor neuron); above it is indicated the upper motor neuron. To the right is the posterior root with its ascending fibre and collateral within the cord (sensory neuron). The parts degenerated are marked with dots. Note that the degeneration in the sensory neuron extends up to the point of entrance of the fibre into the cord.

The chief symptom is ataxia of the limbs which develops gradually. The gait is reeling and there is unsteadiness on standing. The arms are ataxic and fine movements almost impossible. The speech is difficult. The motor power is not diminished. Sensation is not affected. Nystagmus is often present. These symptoms resemble those of Friedreich's disease. But in hereditary cerebellar ataxia the tendon reflexes—knee-jerks—are normal or increased, optic atrophy often occurs, and occasionally the Argyll-Robertson pupil is present. Paralysis of ocular muscles may occur. Scoliosis and the peculiar deformity of the foot (so common in Friedreich's disease) are absent; the onset of the symptoms is usually after the age of 20; and marked mental symptoms (mental weakness and irritability) may develop. The course of the disease is very chronic. Death occurs from some intercurrent affection or from general weakness. Pathologically, atrophy of the cerebellum has been found in some cases but not in all, whilst usually there have been no tracts of degeneration in the cord.

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COMBINED DISEASE OF THE POSTERIOR AND LATERAL COLUMNS OF THE SPINAL CORD

Combined degeneration or disease of the posterior and lateral columns of the spinal cord occurs in the following affections :—

1. In Friedreich's disease.
2. In rare cases of general paralysis of the insane.
3. In rare cases of tabes, degeneration of the crossed pyramidal tracts has been found in addition to the sclerosis of the posterior columns and the other typical tabetic lesions.
4. In primary lateral sclerosis, slight and unimportant changes have been found in the posterior columns, chiefly in the columns of Goll in the cervical region. Also the direct cerebellar tracts have been degenerated in some cases.
5. In amyotrophic lateral sclerosis occasionally there has been a little degeneration in Goll's columns in the cervical region of the cord, in addition to the sclerosis in the lateral columns.

(The four spinal diseases mentioned have been already described.)

6. In addition to these diseases there is an important group of cases, which have been described as *combined degeneration or sclerosis of the posterior and lateral columns.* Sub-varieties of this group are :—
   (*a*) The ataxic paraplegia (of Gowers).
   (*b*) The sub-acute combined degeneration (of Russell, Batten, and Collier).
   (*c*) The diffuse degeneration of the spinal cord (of Putnam).
7. Degeneration of the posterior and lateral columns is sometimes found in (*a*) anæmia (pernicious and other forms), (*b*) pellagra, (*c*) lathyrism, (*d*) ergotism, (*e*) in a few rare cases of leucocythæmia, cancer, Addison's disease, ulcerative endocarditis, etc.
8. In Erb's syphilitic spinal paralysis, the few pathological examinations hitherto published have shown disease of the posterior and lateral columns (*see* p. 393).
9. The spinal sclerosis of senile individuals is an affection in which there is slight postero-lateral sclerosis.

DEGENERATION OF THE POSTERIOR AND LATERAL COLUMNS

In 1877 Kahler and Pick described cases of combined system affection of the cord, afterwards Westphal, Strümpell and Dana recorded similar cases, and now many sub-varieties of the disease are recognised.

The combined degeneration affects the columns of Goll, the crossed pyramidal and the direct cerebellar tracts ; in some cases Burdach's columns are also affected partially, but the posterior root zone, the peripheral portion of Burdach's columns and the fibres adjacent to the commissure often escape. Frequently, however, the degeneration in the white matter is diffuse.

The **Symptoms** vary somewhat with the extent of the degeneration ;

(i.) If the crossed pyramidal tracts are chiefly affected, and the changes in the posterior columns do not extend down to the lumbar region, there is rigidity of the muscles, increased knee-jerks, motor weakness, and in addition ataxia owing to the degeneration in the posterior columns. The symptoms are those of spastic ataxic paraplegia. Other symptoms such as shooting pains, sensory affections, and bladder troubles may or may not develop. (ii.) If the changes in the posterior columns are most marked and extend to the lumbar region, the knee-jerks are absent, and other signs of tabes may be present, but in addition to ataxia there is motor weakness, and Babinski's extensor form of plantar reflex is present—an indication of lesion of the crossed pyramidal tracts.

Not infrequently the symptoms of the first group are gradually succeeded by those of the second group. In other cases ataxia is prominent at an early period, spastic paraplegia later.

The **diagnosis** is often difficult during life. When the changes in the posterior columns are slight, the symptoms thereof may be latent, and the case is usually diagnosed as spastic paraplegia. If in addition to the symptoms of spastic paraplegia there is ataxia, or a history of ataxia at the early part of the illness, this fact would be in favour of combined postero-lateral sclerosis.

Cases of tabes with sclerosis of the lateral columns, or cases of combined sclerosis resembling true tabes more or less, may be distinguished from uncomplicated tabes by the spastic gait, by the paraplegia or paraparesis, and by the extensor type of plantar reflex (Babinski's) which occur in the two former affections.

As regards the **etiology,** in some cases of combined postero-lateral sclerosis, a congenital tendency is probable ; in most cases there is no relation to syphilis, but in some a syphilitic origin is possible. Many cases have been associated with severe or pernicious anæmia, or cachexia from various causes—leucocythæmia, Addison's disease, cancer, septicæmia, ulcerative endocarditis, tuberculosis, etc.

As Marie has pointed out the pathological changes are chiefly localised to the parts of the transverse area of the cord supplied by the posterior arterial system (Fig. 153A) or to the peripheral arterial system. Recently the changes have been attributed to an affection of the lymphatics of the cord.

Ataxic Paraplegia.

This is the name which Sir William Gowers has given to a group of cases of combined postero-lateral sclerosis.[1]

The **etiology** is obscure ; the disease generally begins between the ages of 30 and 40 : a history of syphilis is very rare.

The **symptoms** are usually gradual in onset, and briefly stated are those of spastic paraplegia with ataxia. The legs are first affected. Sometimes the legs only are affected ; in other cases the arms also suffer. There

[1] The account here given is based chiefly on the description of Sir Wm. Gowers.

is unsteadiness in walking, and in standing with the feet together : Romberg's symptom is present; and there is inco-ordination of the movements of the legs when the patient is in bed. There may also be inco-ordination in the movements of the hands. The movements of the legs (especially flexion at the hip and knee) are weak. Dull pain in the sacral region is often an early symptom. Lancinating pains and girdle sensations are almost always absent. Hyperæsthesia and impairment of sensation are usually absent.

The knee-jerks are increased, rectus-clonus and ankle-clonus are usually present. The abdominal and cremasteric reflexes are sometimes absent. The plantar reflex is of the extensor type.

There is no atrophy of the muscles of the limbs. The bladder and rectum are usually not paralysed; but sometimes the sphincters are impaired, and the bladder may be paralysed at a late period. The pupils react to light. Optic atrophy is very rare : there are no signs of affection of the cranial nerves, with the exception of nystagmus on movement of the eyes, which is not infrequently present. As the disease progresses the symptoms resemble more strongly those of spastic paraplegia. Sir William Gowers points out that the symptoms sometimes differ slightly from the description just given. Occasionally sensation is impaired, the knee-jerks are lost, and a girdle sensation is present.

Death occurs from bed-sores, cystitis and its consequences, or from intercurrent disease.

Diagnosis.—The chief symptom at the onset is ataxia ; at a later period the symptoms of spastic paraplegia are more prominent.

From tabes the disease is distinguished by the increase of the knee-jerks, the ankle-clonus, the spastic condition of the legs and loss of power, by the absence of lancinating pains, and by the normal reaction of the pupils to light.

From primary lateral sclerosis it is distinguished by the ataxia.

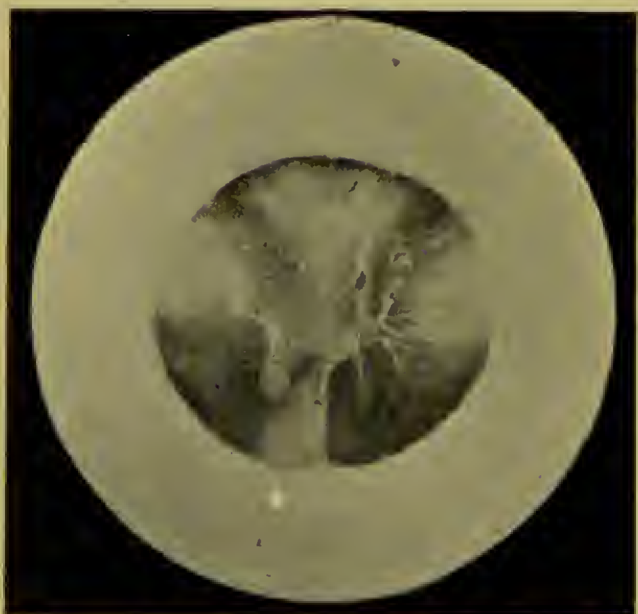
From Friedreich's disease ataxic paraplegia is distinguished by the loss of the knee-jerks, the absence of ankle-clonus and rigidity, the presence of pes cavus and spinal lateral curvature, and the frequent family history of the disease in the former affection.

From cerebellar tumour with ataxia, ataxic paraplegia is separated by the headache, vomiting and optic neuritis and the absence of paresis or paralysis of limbs in the former disease. The more rapid onset of acute and subacute myelitis distinguishes these affections from ataxic paraplegia.

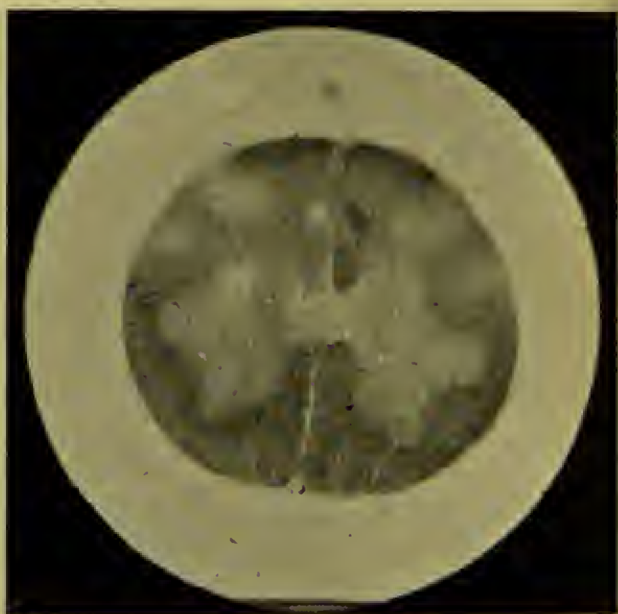
Pathological Anatomy.—Examination of the spinal cord reveals sclerosis of the posterior and lateral columns. The extent of the sclerosis varies considerably in different cases. The sclerosis is usually less intense in the lumbar than in the dorsal region. In the lower and mid-lumbar regions the posterior columns are sometimes unaffected, whilst in the dorsal and dorso-lumbar regions they are markedly affected. The sclerosis in the posterior columns affects both the columns of Goll and Burdach, but it has not the special intensity in the root zone which is character-

istic of tabes. The posterior columns, adjacent to the grey commissure and neck of the posterior horns, are usually unaffected. Sometimes the degeneration does not extend up to the posterior surface of the cord, only the middle three-fifths of the posterior columns being affected. When the degeneration in the lower part of the cord is considerable,

A



B



C

FIG. 155.—Spinal Cord; Ataxic paraplegia. Pal's stain.

A = Cervical Region. Note degeneration, pale area, in lateral columns, in region of anterior pyramidal tracts and in posterior columns: but the posterior columns are not affected close to the posterior grey matter.

B = Dorsal Region.

C = Lumbar Region. In the lumbar region there is degeneration of each crossed pyramidal tract and a small oval area of degeneration in the posterior columns on each side of, and a short distance from, the posterior median septum.

there is marked secondary degeneration in Goll's columns in the upper part.

In the lateral columns the crossed pyramidal tracts are chiefly affected, but the degeneration often extends into the white matter in front of this tract, sometimes into the lateral limiting layer and the direct cerebellar tract. The anterior pyramidal tract may also be degenerated, and occasionally the whole of the peripheral white matter of the antero-lateral columns, as well as the crossed pyramidal tracts, is affected. The posterior nerve roots, the grey matter, and the meninges are normal (*see* Figs. 155 and 156).

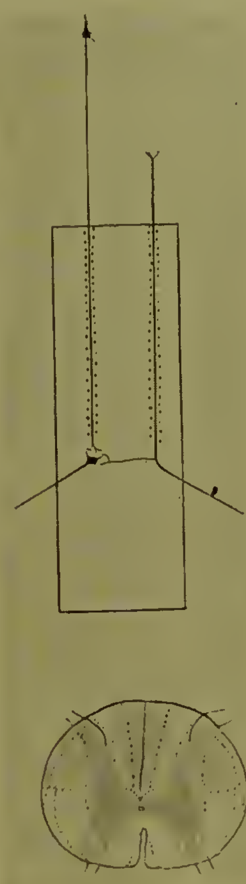


FIG. 156.—Postero-lateral Sclerosis (Ataxic paraplegia, etc.). Lower figure = transverse section of cord; degenerated parts shaded. Upper figure = longitudinal section. To the left are indicated a fibre of the anterior root and its cell of origin, lower motor neuron; and above it the upper motor neuron. To the right is the posterior root and its ascending fibre and collateral within the cord (sensory neuron). The parts degenerated are marked with dots. Fibres of the posterior root not degenerated for a short distance after entering the cord.

The ataxia is probably due to the disease of the posterior columns, the spastic paraplegia to the affection of the crossed pyramidal tracts, and the persistence of the knee-jerks to the fact that the posterior root zone is either not invaded, or is little affected by the disease. This is a point of difference between the lesions in ataxic paraplegia and Friedreich's disease.

SUB-ACUTE DEGENERATION OF THE SPINAL CORD.[1]

Under this name Risien Russell, Batten, Collier, and others have carefully described a form of postero-lateral degeneration closely allied to the affection just considered, if it is not actually the same disease. But as there are minor points of difference, it has appeared advisable to give a short account of the group of cases recorded by these authors in a separate section.

Symptoms.—*First stage.*—The onset is usually gradual. Numbness and tingling in the legs are the first symptoms. Later slight spastic paraplegia and slight ataxia develop. In some cases the spastic symptoms become more marked, in others the ataxic. The subjective sensations in the legs increase; the deep reflexes become exaggerated,

[1] This account is based on the description of Russell, Batten, and Collier.

and Babinski's extensor form of plantar reflex is obtained. This is the longest stage of the disease.

Second stage.—The transition from the first to the second stage is abrupt. The ataxia increases so that the patient is unable to stand. Marked anæsthesia develops rapidly in the legs and trunk. Paresis gradually develops into marked spastic paraplegia. Girdle sensations and shooting pains are often present. The knee-jerks are increased; ankle-clonus and Babinski's extensor type of plantar reflex are obtained. The sphincters are not affected.

Third stage.—The transition from the second to the third stage is rapid. Complete flaccid paraplegia develops. There is rapid wasting of muscle with loss of faradic excitability, whilst galvanic excitability is diminished with or without polar change. The knee-jerks and ankle-clonus disappear. But Babinski's extensor type of plantar reflex is present and the superficial reflexes are increased. The anæsthesia becomes absolute. Œdema of the legs and trunk often develops, and there is frequently slight irregular pyrexia. There is absolute incontinence of both sphincters. Subjective sensory symptoms may develop in the arms along with slight rigidity or ataxia. The average duration of the disease is under nine months.

Diagnosis.—At the early stage, the disease is distinguished from disseminated sclerosis by the average age of onset—forty (i.e. a little later than that of disseminated sclerosis), by the absence of nystagmus, by the irregular pyrexia, by the symmetrical affection of all the limbs with slight ataxia and spastic paresis, the last mentioned symptoms being most marked in the legs.

At a later period, second stage, the disease is distinguished from other forms of paraplegia by the history that at first ataxia was more marked than the paresis.

At the third stage the disease is distinguished from tabes by the history of spastic paralysis changing to flaccid paralysis, by the complete paralysis and anæsthesia, by the slight pyrexia and œdema of the legs, by the atrophy of muscle and loss of faradic excitability, by the presence of Babinski's extensor type of plantar reflex, and the absence of the Argyll-Robertson pupil.

From peripheral neuritis the disease is distinguished by the history of the spastic condition of the legs at the onset, by the extensor type of plantar reflex and the affection of the sphincters in the sub-acute degeneration of the cord.

Pathology.—The changes are most marked in the mid-dorsal region of the cord, and in an advanced case the degeneration affects almost the whole of the peripheral white matter; only the grey matter and a narrow strip of white matter around it being unaffected.

Above the mid-dorsal region the degeneration is less extensive and tends to be limited to the columns of Goll and Burdach, the direct cerebellar tracts, the tracts of Gowers and the crossed pyramidal tracts;

often the direct pyramidal tract is degenerated. The degeneration can be traced into the medulla and pons, but the upper part of the pons and the brain are normal.

In the lower part of the cord the changes diminish, and consist chiefly of degeneration of the crossed pyramidal tracts, with patches of degeneration in the posterior columns. The grey matter, Lissauer's columns, the posterior nerve roots, the posterior ganglia and the peripheral nerves are usually normal.

The changes consist of two processes—(1) focal destructive lesions, and (2) systemic lesions. These two lesions are blended together. In the focal lesions the changes commence with swelling of the medullary sheath of the nerve fibres, whilst the axis cylinders are unaltered. The swollen sheaths degenerate, and the degeneration products are absorbed; the axis cylinders then disappear and spaces are left in the white matter. These spaces often become large by the fusion of several adjacent ones. At a later stage the neuroglia connective tissue is increased, and sclerosis is produced.

The systemic lesions consist of degenerative changes, similar to those of the secondary degeneration (sclerosis), which follows transverse lesions of the cord. The nerve fibres of the affected tracts have degenerated and the connective tissue is increased.

At the region of the cord most affected, the degenerated area is that supplied by the peripheral spinal vessels (*see* p. 25 and Fig. 26).

In some of the cases no anæmia was present throughout the whole course of the disease; in others it occurred late, along with wasting; whilst in others it was marked from the first, and preceded the nervous symptoms. The blood changes found were those of secondary anæmia and did not correspond to pernicious anæmia.

The cord changes are very symmetrical, and Russell, Batten and Collier think they are probably due to a toxin, which is especially prone to affect certain parts of the nervous system.

Diffuse Degeneration of the Spinal Cord.[1]

In 1891 Putnam drew attention to a group of cases of diffuse degeneration of the spinal cord, "occurring in enfeebled persons past middle life and especially in women." Recently Putnam and Taylor have more fully described this group of cases.

The symptoms appear to correspond to those described by Russell, Batten, and Collier. Most of Putnam's cases were over fifty years of age. Syphilis was a rare antecedent. The patients were in feeble health or were of advanced age. Seven out of fifty cases suffered from severe anæmia.

Pathology.—The chief pathological features of this group of cases were :—

[1] This account is based on the description of Putnam and Taylor.

1. A diffuse degeneration, for the most part limited to the cord, often in more or less discrete patches.

2. A constant involvement of the posterior and lateral columns, without strict regard to neuron systems.

3. Predominance of the lesions in the cervical and dorsal regions.

4. The freedom from degeneration of nerve roots (motor and sensory), peripheral nerves and grey matter.

5. Insignificant vascular changes.

The occurrence of degeneration in discrete areas, varying from segment to segment, is particularly characteristic.

The nerve fibres of the affected areas are at first swollen. The myelin sheaths are dilated, and present a vacuolated appearance, whilst often the axis cylinders are relatively normal. Later the whole fibres may degenerate.

In some cases there is a neuroglial sclerosis in addition, but often there is little tendency to neuroglial proliferation. The distribution of the lesion is apparently dependent on the anatomical distribution of the pial or peripheral vessels of the cord (*see* p. 25). Under the influence of an unknown cause (probably a toxin), those areas of the cord suffer first which are least supplied with blood in the normal condition.

Putnam and Taylor conclude that well defined lesions of the spinal cord exist which may, for the present, be termed simply " diffuse degeneration " ; that the anæmic states are at times concomitant, but are not necessarily causes. The actual causes of the disease are still obscure.

Changes in the Spinal Cord associated with Pernicious and Severe Anæmia.

In 1887 Lichtheim drew attention to changes in the spinal cord associated with severe anæmia ; afterwards they were carefully described by Minnich, Nonne, Bowman, Michell Clarke, James Taylor, F. Billings, and others. The cord changes have been found not only in true pernicious anæmia, but also in severe anæmia due to the bothriocephalus latus, anchylostoma duodenale, and to other causes.

Symptoms of affection of the spinal cord occur only in a small percentage of cases of pernicious or severe anæmia. They may occur early or late. The pathological changes in the cord are sometimes marked when the symptoms are very slight, and vice versa ; also changes are sometimes found on examination of the cord in anæmia and yet there have been no symptoms during life. Often the nervous symptoms are chiefly subjective, consisting of numbness and tingling of the legs and arms ; in these cases the knee-jerks may be absent. In other cases the subjective symptoms are associated with unsteadiness in walking or ataxia ; paresis and pains in the legs, and objective sensory symptoms may develop. The arms may be affected early. The knee-jerks may be normal, absent, or increased. The legs may become spastic : ankle-clonus and the extensor form of plantar reflex may develop. There are

no pupillary changes. At a later date a complete flaccid paraplegia, with loss of control over the bladder and rectum, may develop. Anæmia, severe or pernicious, with impairment of health is present.

The affection runs a comparatively rapid course and terminates fatally in a few months or a year, though remissions sometimes occur. Death is due to asthenia, bed-sores, or cystitis. The prognosis is very unfavourable.

Fig. 157.—Degeneration in the Posterior Columns of Spinal Cord (pale area); pernicious anæmia. Pal's stain. Lateral columns not affected.

Pathological Anatomy.—The spinal changes found in pernicious anæmia are of two kinds—(1) small foci of sclerosis and capillary hæmorrhages, which tend to become confluent; (2) diffuse degeneration. The latter is the condition usually associated with the spinal symptoms.

In many cases the pathological changes in the spinal cord affect the posterior columns only; in other cases both posterior and lateral columns are degenerated. The changes are most marked, as a rule, in the upper dorsal and lower cervical regions; they gradually diminish below the regions mentioned, and may disappear in the lower lumbar region; they also diminish when traced upward. When changes are present in the lateral columns they affect the crossed pyramidal tracts and often the adjacent white matter, as in ataxic paraplegia. The anterior pyramidal tracts may be involved at different levels, but not always symmetrically.

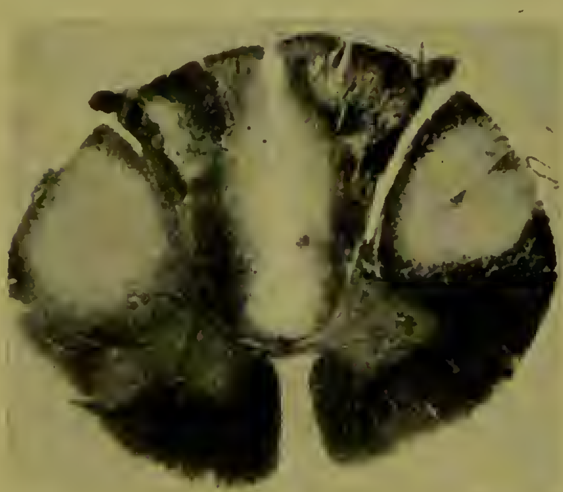
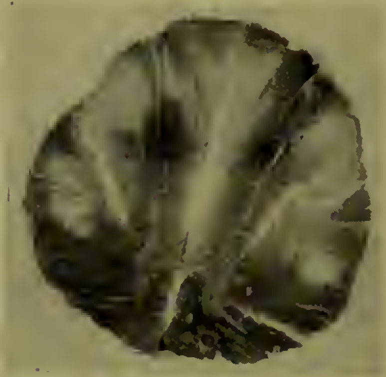


Fig. 158.—Microphotograph (× 4 diameters) of Cervical Region of Cord. Weigert-Pal's stain. Pale areas = degeneration. Pernicious anæmia.

The lateral degenerated area may be separated from the surface of the cord in the cervical and upper dorsal regions by a narrow zone of normal or only slightly degenerated tissue. Often the degeneration is not strictly limited to the crossed pyramidal tract in the cervical and dorsal regions. Small round patches of degeneration may be seen in the white matter just in front of the crossed pyramidal tract (Fig. 159). Also in many of the sections of the dorsal and cervical cord, the degener-

ated area in the lateral column has not exactly the shape and size of the crossed pyramidal tract, and sometimes does not affect the whole of the tract. The degeneration often appears as round patches or streaks



FIGS. 159 and 160.—Dorsal Regions (From same case as Fig. 158).

surrounding the blood-vessels. In the lumbar and sacral regions the lateral degeneration may be localised to the crossed pyramidal tracts, and have the size and shape of these tracts (Figs. 158–161).

In the posterior columns the white matter adjacent to the posterior grey horns and commissure (i.e. the posterior root zones and the posterior ventral fields) is usually not degenerated. Lissauer's zone is usually unaffected ; and the posterior roots are normal or only slightly affected.

When the changes in the posterior columns are extensive there is, in the cervical region, marked degeneration of Goll's columns, and sometimes irregular patches of degeneration in Burdach's columns. The most posterior part of Goll's columns, adjacent to the surface of the cord, may be less degenerated than other parts of these columns.

FIG. 161.—Lumbar Region (From same case as Figs. 158–160).

Evidently the changes in Goll's columns in the cervical region are chiefly secondary to changes in the posterior columns of the cord at a lower level.

In the cervical region, often the secondary degeneration in Goll's columns is separated by a narrow streak from changes of a primary nature around the intermediate septum. The latter do not reach to the surface of the cord.

In the lower dorsal, lumbar and sacral regions the changes in the posterior columns gradually diminish from above downwards and may be chiefly along the course of the posterior intermediate septal artery. The degenerated patches are usually separated from the posterior median fissure by normal fibres (*see* Figs. 160 and 161).

When only the posterior columns are affected the changes are usually most marked in the cervical region. The situation is seen in Fig. 157. As in ataxic paraplegia only the median three-fifths of the columns of Goll may present degenerative changes.

The posterior ganglia have been found unaltered. The grey matter and nerve cells of the anterior horns are unaffected as a rule. The cells of the brain cortex and the peripheral nerves have usually been found normal. The degeneration cannot be called systemic, because

FIG. 162.—Microphotograph of White Matter of Lateral Column. At upper part (paler portion) there is marked degeneration of nerve fibres, and large spaces are left from which the nerve fibres have disappeared, causing a vacuolated appearance (Weigert-Pal's stain).

an entire system is not involved, nor is a whole neuron implicated in the degenerative process.

Homén points out that the changes are usually in relation to the septa and vessels.

The vessels often present changes, especially in the older foci of degeneration. The perivascular sheaths are dilated, and often contain detritus, granules, leucocytes and compound granular cells. The vessel walls, and not infrequently the neuroglia around, are thickened in advanced cases. The external coats of the vessels sometimes show cellular infiltrations. The small vessels often present hyaline degeneration, and the lumen is narrowed; in rare instances they are obliterated or thrombosed.

The first changes in the spinal degeneration are usually in the form

of scattered spots or streaks, near a septum or a vessel with altered walls; and not infrequently they are scattered around the terminal branches of a vessel or the terminal fibres of a septum.

The alteration in the nerve elements consists at first of scattered degeneration of fibres. The myelin sheath becomes swollen, and then degenerates. The axis cylinder is usually swollen also and the whole fibre may present a homogeneous or granular appearance. In a more advanced stage, large spaces are seen from which nerve fibres have dis-

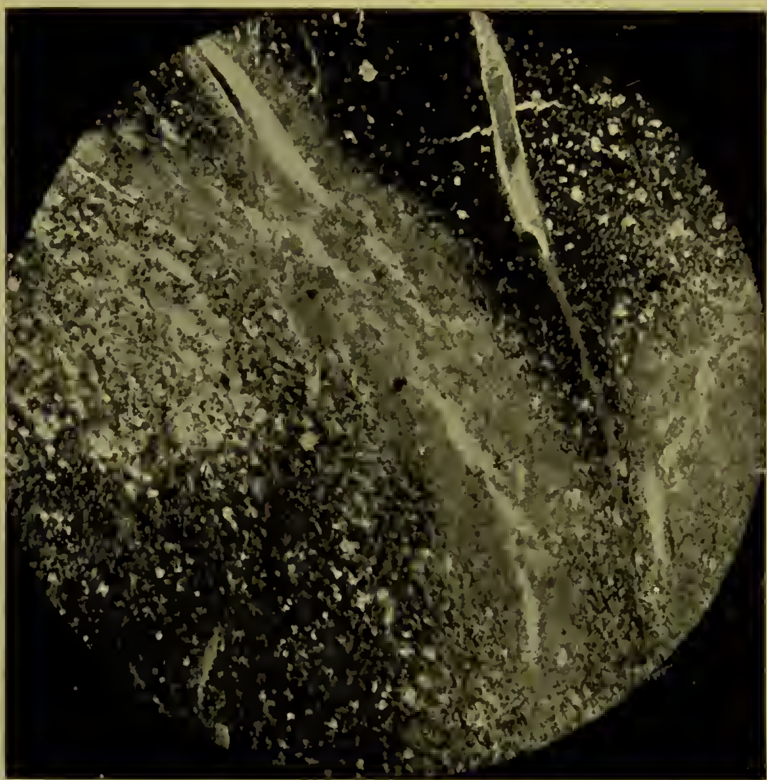


FIG. 163.—Microphotograph of Posterior Columns, showing streak of degeneration (pale) in the course of the posterior intermediate septal artery (this degenerated pale area runs obliquely through the centre of the photograph). To the right of the degeneration is the posterior median fissure.

appeared entirely, and thus a vacuolated appearance is produced. Sometimes these spaces are very large and equal to the space which would be occupied by many nerve fibres.

Secondary sclerosis may occur with increase of neuroglia.

Two kinds of degeneration have been described. In one, which is more acute, the nerve fibres break down into a *debris* and fat droplets. This process is not usually attended with an increase of neuroglia, and therefore, when the products of the degenerated nerve fibres are absorbed a vacuolated appearance is left. In another form of the degenerative process the changes are more chronic, the neuroglia is increased, and sclerosis is produced.

When the spinal changes are well marked the degeneration in the crossed pyramidal tracts of the lumbo-sacral region and in Goll's columns

of the cervical regions appears to be of the nature of secondary degeneration. But in other parts, i.e. in the posterior columns of the dorsal and lumbo-sacral regions, and in the lateral columns of the cervical and dorsal regions, the degeneration is often clearly related to the course of blood-vessels. In these columns, round patches of degeneration may be seen with one or more greatly dilated vessels in the centre; or streaks of degeneration follow the course of a dilated vessel. In the posterior columns the degeneration, when slight, may be clearly localised around the course of the posterior intermediate septal arteries (*see* Fig. 163).

The changes are often clearly localised to the region supplied by the posterior arterial system of spinal arteries, i.e. to the shaded area in Fig. 153A.

In four cases of spinal lesions in anæmia in which I have examined the spinal cord pathologically, the posterior columns only were affected in three; in one both posterior and lateral columns presented degeneration.

Figs. 158 to 161 show the small patches of degeneration in the posterior and lateral columns in the last mentioned case.

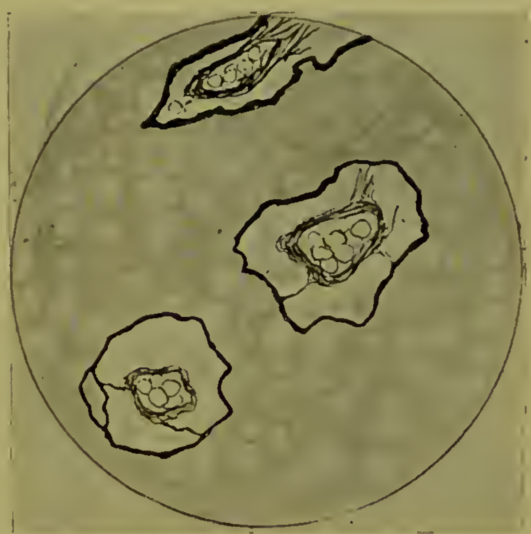


FIG. 164.—Blood Vessel of the Cord in Pernicious Anæmia, with spinal postero-lateral degeneration (van Gieson's stain). Note adventitial lymph sheath much distended. Walls of vessel thickened.

In all of the three cases in which the posterior columns only were affected the degenerated areas were small, they did not extend up to the surface of the cord, nor into the posterior root zone. The degenerated patches were clearly localised around the course of the posterior intermediate septal artery, or to the inner side thereof—between the artery and the posterior median fissure.

All the degenerated patches showed the peculiar vacuolated appearance already described.

The diffuse spinal cord lesions, reported by Putnam, by Russell, Batten and Collier, and others, and the changes in ataxic paraplegia (as described by Sir William Gowers) do not differ essentially from those found in pernicious anæmia.

Some of the cases described as ataxic paraplegia, or diffuse or combined spinal degeneration have been associated with pernicious or severe anæmia, and probably the spinal changes have sometimes been due to the anæmia. But this has not always been the case.

In fifty of the cases of combined postero-lateral spinal degeneration reported by Putnam and Taylor seven suffered from pernicious anæmia.

In the nine cases reported by Russell, Batten and Colier two only suffered from pernicious anæmia; also in nine reported by Burr two only suffered from pernicious anæmia.

F. Billings draws the following conclusions from the study of the spinal changes in severe anæmia:—

1. That there is a well-established relation of diffuse cord degeneration with pernicious anæmia.

2. It seems highly probable that the hæmolysis and cord changes are due to the same toxin.

3. While the source of the toxin is unknown, the fact that gastro-intestinal disturbance is so common in the disease would lead one to suppose that it is of intestinal origin.

4. The diffuse degenerations of the spinal cord, which occur in conditions without pernicious anæmia, do not appear to differ essentially from those of pernicious anæmia.

5. It is possible that a common blood circulating poison exists which may expend its force upon the blood in one individual, upon the nervous apparatus in another, and coincidentally upon the blood and spinal cord in others.

Diagnosis.—These cases of spinal degeneration in anæmia differ from ataxic paraplegia by the presence of severe anæmia. They differ from tabes by their more rapid course, by the absence of pupillary and trophic changes, and by the presence of severe anæmia. The Babinski extensor form of plantar reflex occurs when the crossed pyramidal tracts are diseased in the cord lesions of severe anæmia, and this sign is of much importance in distinguishing the affection from true tabes. From disseminated sclerosis the diagnosis may be difficult, but intention tremor, nystagmus, and scanning speech, if present, would point to this affection.

The **treatment** of the various forms of postero-lateral sclerosis is unsatisfactory. If anæmia be present iron or arsenic, and the treatment suitable for pernicious or severe anæmia, are indicated. General tonic treatment is suitable when malnutrition is present. Complications should be treated as in cases of myelitis. Putnam has found Frenkel's treatment—re-education of muscular movements—of great importance in diminishing the ataxia. For the pains in the limbs the drugs recommended in the treatment of tabetic pains may be used.

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Pellagra.

This is an epidemic disease, met with in Italy, in which sclerosis of the lateral and posterior columns of the cord has been found on pathological examination (Tuczek). The sclerosis has a similar distribution to that in other forms of postero-lateral sclerosis. Changes are almost constantly present in the posterior columns, and usually in the lateral pyramidal tracts. According to some observers the changes in the posterior columns have affected chiefly the endogenous fibres. Other observers have recorded changes in the posterior root fibres. Also there is often atrophy of the nerve cells of the anterior horns of grey matter, and chronic inflammatory changes in the pia mater have been recorded. The disease is due to a poison taken into the system through the consumption of diseased or unripe maize.

Fig. 165.—Spinal Cord; pellagra. Degenerated parts of white matter shaded (diagrammatic).

Briefly stated, the symptoms consist of weakness of the legs, passing on to spastic paresis, with increase of the reflexes, and unsteadiness in the movements. Sensation may be normal or diminished, or hyperæsthesia may be present. The disease has generally a chronic course; anæmia and cachexia are usually present. (Sometimes acute symptoms occur resembling those of spinal meningitis, and post mortem examination reveals changes resembling those of myelitis along with meningitis.)

Lathyrism is the disease caused by chronic poisoning by the meal prepared from lathyrus seeds, which is used for the preparation of bread in Algiers and India and other countries. The nervous symptoms have been those of spastic paraplegia or of combined sclerosis, and are probably due to degeneration of the lateral pyramidal tracts of the cord.

Ergotism.—In an epidemic of ergotism Tuczek observed symptoms resembling, in some degree, those of tabes. Mental symptoms—mania, melancholia, stupor, and epileptic attacks—are also often present. Usually there are indications of cachexia. The knee-jerks are lost, and ataxia, paræsthesia, analgesia, Romberg's symptom and shooting pains are present. There is a tendency to improvement. Changes are found in the posterior columns, chiefly in Burdach's columns, whilst Goll's columns are spared.

The changes are not progressive and may subside. They affect chiefly the intra-medullary fibres of the posterior roots (but the extra-medullary part may also be affected). The degeneration is usually more acute than in tabes.

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Spinal Changes in Arterio-Sclerosis (Senile Paraplegia).

Senile changes occur in the spinal cord as in the brain, but gross pathological lesions, or marked symptoms, are seldom produced by the spinal changes. The blood-vessels of the spinal cord in old age may present the changes of arterio-sclerosis, but softening or hæmorrhage in the cord substance, as a result thereof, is exceedingly rare. Other senile changes in the spinal cord are occlusion of the central canal, thickening of the meninges, marked pigmentation and slight atrophy of the ganglion cells of the anterior horns, and the presence of numerous corpora amylacea.

Sir William Gowers has described a rare condition, to which he gave the name of simple senile paraplegia. It occurs in patients over fifty years of age, and is characterized by weakness of the legs with slowness of movements. There is no wasting and no affection of sensation. The knee-jerks are normal and ankle-clonus is absent. The onset is gradual and the symptoms slowly progressive, but the loss of power is seldom so great as to prevent the patient standing. The condition of the legs somewhat resembles that in paralysis agitans, and Sir William Gowers thinks that the affection is probably a partial development of the morbid process of that disease—possibly owing to changes affecting only the motor nerve cells of the leg centres of the brain cortex. Similar cases are also met with in which the reflexes are increased (*Diseases of the Nervous System* : Gowers and Taylor, 3rd edition, Vol. 1. 1899, p. 530).

Demange has recorded cases of spastic paraplegia, due to atheroma of the spinal vessels, associated with perivascular sclerosis and streaks of diffuse sclerosis (*Revue de Médecine*, 1885, p. 1).

Oppenheim has described senile paraplegia caused by arterio-sclerosis of the spinal vessels, and perivascular sclerosis affecting chiefly the white matter of the cord. The symptoms are those of spastic paraparesis. Affection of sensation and of the sphincters is rare.

The spinal changes in arterio-sclerosis have been carefully studied by Pic and Bonnamour (*Revue de Médicine*, 1904, Nos. 1 and 2). They record the pathological examination in seven cases. The spinal cord did not present any definite tracts of sclerosis, but the spinal arteries were thickened, there was perivascular sclerosis and a diffuse sclerosis extending from the peri-arterial changes.

Pic and Bonnamour conclude that in old people suffering from arterio-sclerosis there is an affection characterized by feebleness of the legs, increase of the knee-jerks, and slowness of gait. The affection develops slowly but progressively. The autopsy reveals general atheroma, and around the arteries of the cord a diffuse spinal sclerosis, not systemic, but with a marked predominance in the crossed pyramidal tracts and in the posterior columns. It is probable, but not clearly proved, that this diffuse interstitial myelitis is of vascular origin. Instead of being

generalised and diffuse, the spinal sclerosis may be localised and produce symptoms resembling those of sclerosis of the spinal tracts.

THE SYMPTOMS DUE TO PERIPHERAL NEURITIS OR SPINAL LESIONS IN DIABETES MELLITUS

In many cases of diabetes mellitus, nervous symptoms occur which are due to changes either in the peripheral nerves, or in the fibres of the posterior nerve roots just after they have entered the spinal cord. There are several clinical forms, but the feet and legs are most frequently affected.

The chief symptoms of these cases of so-called diabetic neuritis are :—

1. Subjective symptoms in the feet and legs : pain and burning sensation ; tenderness and hyperæsthesia of skin and muscles ; cramps in the calf muscles. The symptoms are often slight, but the pain and tenderness are sometimes so severe as to prevent sleep, and the patient is unable to bear the contact of the bed clothes with the legs.

2. Loss of the vibrating sensation on the soles of the feet and big toe nails, or on these parts and on the malleoli and tibiæ (*see* p. 80).

3. Loss of the tendo Achillis reflexes ; the knee-jerks being present in some cases, lost in others.

In the majority of cases pain, tenderness, and hyperæsthesia are the prominent symptoms. As Leyden has pointed out, the so-called neuritis of diabetes is usually of the hyperæsthetic form. Though there may or may not be a sensation of numbness and tingling, nevertheless on objective examination usually no loss of sensation to tactile impressions, pain or temperature can be detected ; and usually *the only objective sensory defect is loss of the vibrating sensation*, as tested by a vibrating tuning-fork.

Dropped feet and true paresis or paralysis, with or without anæsthesia to tactile sensation, pain, and temperature—symptoms resembling those of a well marked alcoholic neuritis—are occasionally observed, but they are very rare.

Perforating ulcers like those of tabes occasionally occur, and often in these cases there are pains in the legs and loss of the tendo Achillis jerks, or of both the tendo Achillis jerks and knee-jerks. In rare cases the pains and other symptoms are in the distribution of the anterior crural nerves or in the arms.

The tendo Achillis reflex is often lost in diabetes ; in 50 consecutive cases of diabetes (chiefly private patients) I found the Achillis jerks both lost in 19, present in 29, one absent and one present in 2.

The knee-jerk is also often lost ; in 100 cases of diabetes, chiefly hospital patients, I found the knee-jerks both lost in 49, one lost and one present in 6, both present in 45. But in 50 private patients I found the knee-jerks both lost in 6 only ; one lost, one present, in 1 case ; both present in 43.

As in tabes, the Achillis jerks are usually lost in diabetes before the knee-jerks. When the Achillis jerks are lost the knee-jerks may be lost or present, but when the Achillis jerks are present the knee-jerks are nearly always present. Thus, in the 50 cases of diabetes already mentioned, the Achillis jerks were both lost in 19, but the knee-jerks were lost in only 8 of these cases.

The vibrating sensation is lost in the legs (at some part where it is felt normally) in many cases of diabetes, although other nervous symptoms may be very slight.

Resemblance of the Symptoms to those of Tabes.—From the description just given, it will be evident that many symptoms which occur in tabes may be met with amongst the nervous complications of diabetes, as, for example, the loss of knee-jerks and of tendo Achillis reflexes, perforating ulcers, pains in the legs, impairment of sensation, and sexual impotence. In diabetes, as in tabes, the tendo Achillis reflex is usually lost before the knee-jerk; in both affections, when these two deep reflexes are lost, the abdominal and epigastric reflexes are very often increased. The pains in the legs in diabetes are usually of a dull, aching or gnawing character, and the typical shooting pains of tabes are usually absent; but in a few cases which have come under my observation, the description of the pains given by the diabetic patient has corresponded to that of the sharp, stabbing, momentary pains of tabes. In diabetes, as in tabes, very often the first objective sensory symptom is loss of sensation to the vibrating tuning-fork, whilst other forms of sensation have not been lost.

The diagnosis from true tabes is, however, usually very easy. The difficulty occurs chiefly in cases in which the thirst and diuresis have been absent or slight, and examination of the urine has not been made. The patient has often sought advice on account of pain in the legs, or on account of a perforating ulcer; examination has revealed loss of knee-jerks and of the tendo Achillis reflexes, and a diagnosis of tabes has therefore seemed probable. Hence it is a good practical rule never to omit the examination of the urine for sugar in all cases of severe pains in the legs, in sciatica, in cases in which the knee-jerks or Achillis reflexes are absent, and in cases in which the symptoms of multiple peripheral neuritis are present.

The urinary condition is usually diagnostic. There are, however, several points of difference as regards the nervous symptoms. The usual difference with respect to the pain has been already referred to. Hyperæsthesia of the muscles, tenderness of the calf muscles, is very common in the diabetic nervous complications, whilst this tenderness is usually absent in tabes. In fact, in tabes very often there is muscular analgesia. The deepest pressure of the calf muscles in tabes often fails to produce any pain, whilst in the diabetic nervous complications slight pressure usually causes great pain. Girdle sensations and zones of diminished cutaneous sensation round the chest or abdomen are common

in tabes, but do not occur in diabetes. The Argyll-Robertson pupil, gastric and other visceral crises, Charcot's joint disease, bladder symptoms, muscular hypotonus (in absence of paresis) occur in tabes, but not in the diabetic nervous affections. Primary optic atrophy occurs in 10 to 15 per cent. of the cases of tabes. I have examined the optic discs of a very large number of cases of diabetes, but have never met with a single case of optic atrophy. I am inclined to think that in the few cases on record in which optic atrophy has been found in diabetes, the atrophy has been due to some accidental complication.

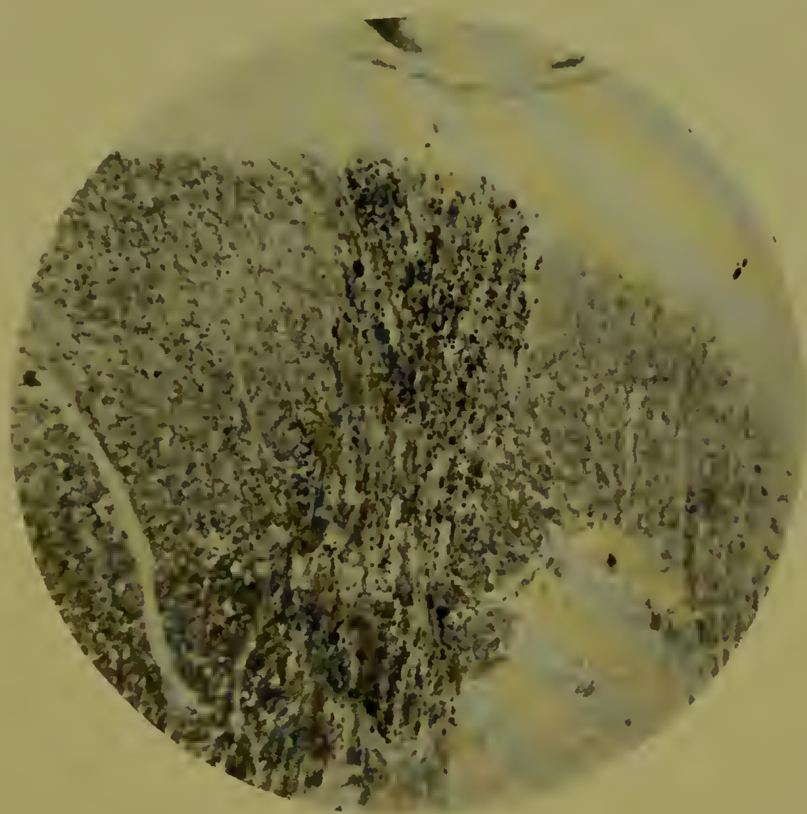

FIG. 166.—Photograph of Section of Posterior Part of Spinal Cord, at entrance of posterior root in severe diabetes mellitus. Degeneration of fibres of posterior nerve root, between surface of Cord and posterior grey matter (pale area at right side of lower part of photograph). Marchi's stain. Posterior root fibres presenting black dots are degenerated (seen in middle of photograph).

Pathology.—The slight nervous symptoms may occur in any form of diabetes, but the more severe nervous symptoms are met with chiefly in the chronic forms and sometimes in the mild cases; usually, but not always, the patients are over forty years of age. The severe nervous complications are very rare in young patients suffering from an acute and severe form of the disease.

From the similarity of many of the symptoms to those of peripheral neuritis, due to "alcoholism" and other causes, the nervous affections in the limbs in diabetes have been attributed to peripheral neuritis. This is certainly the cause of the severe nervous symptoms in many cases. In a number of cases pathological examination has revealed a degenerative

or parenchymatous neuritis in the nerves of the limbs affected. In other cases in which there have been simply loss of knee-jerks and slight pains in the legs, I have not found any changes in the small peripheral nerves of the legs ; a more thorough examination, however, of the *most minute* intra-muscular nerve fibres might possibly have revealed slight degeneration.

But it is possible that the loss of knee-jerks and Achillis jerks and the nervous symptoms may sometimes be due to degeneration in the spinal cord affecting the intra-medullary fibres of the posterior nerve roots ; or at least these changes, as well as changes in the peripheral nerves, may be the cause of nervous symptoms.

In several cases of diabetes I have found changes in the intra-medullary fibres of the posterior nerve roots and in the posterior columns of the cord. Similar changes in the posterior columns have been reported by others. They are probably due to the toxic condition of the diabetic blood.

Fig. 167.—Diabetes Mellitus. Diagram showing position of degeneration (represented by black dots) in Goll's columns in the cervical region of the Spinal Cord. Marchi's stain.

The following is a summary of the spinal changes :—

In sections stained according to Marchi's method, numerous degenerated fibres, stained black, were seen in Goll's columns in the cervical region ; and a few scattered degenerated fibres were seen both in Goll's and Burdach's columns in the dorsal and lumbar regions.

Degeneration, indicated by black dots, was also seen in the fibres of the posterior nerve roots in their intra-medullary course especially between the posterior surface of the cord and the posterior horn of grey matter. The degeneration affected the median bundle of the posterior root fibres, and commenced directly these fibres had passed through the spinal pia mater. The degeneration of the posterior root fibres was best seen in the lumbar and cervical regions. External to the cord (external to the pia mater) the posterior roots were usually quite normal. (In a few sections there was degeneration of the posterior root fibres, just outside the pia mater, but only for a very short distance.) Lissauer's zone was not affected. These changes in the posterior root fibres had the same localisation as the degeneration of the intra-medullary fibres of the posterior roots seen in early tabes and in certain cases of intra-cranial tumour (*see* p. 37).

The changes in the posterior columns are evidently, in some cases, secondary to the degeneration of the intra-medullary fibres of the posterior nerve roots, i.e. they are of the nature of ascending degeneration. As already mentioned, sections stained according to Marchi's method revealed degenerated fibres in the posterior columns, which were most numerous in Goll's columns of the cervical region. Also in the columns

of Goll in the cervical region, aniline blue black, van Gieson's stain, and Weigert's stain revealed slight increase of the neuroglia. The nerve fibres of Goll's columns were mostly diminished in size, but close

FIG. 168.—Diabetes Mellitus. Transverse section of Goll's columns of cervical region of the Spinal Cord (high power of microscope). Numerous degenerated fibres (black dots). Marchi's stain. At each side is Burdach's column in which degenerated fibres are absent.

FIG. 169.—Diabetes Mellitus. Degeneration of intra-medullary fibres of posterior nerve root between the surface of the Cord and the posterior horn of grey matter. Degenerated fibres = black dots. Marchi's stain. Lumbar region of Cord. Extra-medullary portion of posterior root is normal (seen at upper part of figure).

to the posterior median septum some fibres presented swollen axis-cylinders and distended myelin sheaths.

In some specimens changes were well seen by the naked eye on transverse section of the cord hardened in Müller's fluid. The affected parts of the posterior columns of the cord were paler than the rest of the white

FIG. 170.—Diabetes Mellitus. Naked-eye appearances of the Spinal Cord on section, after hardening in Müller's fluid. The normal "white matter" is shaded. The pale portions of the posterior columns are degenerated. The figure to the left = upper cervical region; middle figure = lower cervical region; figure to the right = dorsal region.

matter. In the dorsal region both Burdach's and Goll's columns were affected (pale), whilst in the cervical region Goll's columns only were pale.

In these cases the microscopical changes were much less distinct than those seen by the naked eye.

Treatment.—Often the nervous symptoms here described can be

greatly relieved by medical treatment. It is important to give a careful dietetic and medical treatment suitable to the form of diabetes from which the patient is suffering. For the relief of the pains the following drugs are very useful—aspirin, gr. 10 or 15 three or four times a day; sodium salicylate, gr. 10 or 15 three or four times a day; antipyrin, gr. 10 three times a day. Extract of belladonna with glycerine (equal

Fig. 171.—Diabetes Mellitus (second case). Naked-eye appearances of the Spinal Cord on section, after hardening in Müller's fluid. The normal "white matter" is shaded. The pale portions of the posterior columns are degenerated. Figure to the left = cervical region; middle figure = dorsal region; figure to the right = upper lumbar region.

parts) or cocaine ointment (B.P.), applied to the painful and tender parts is of service.

Diabetes associated with Diseases of the Spinal Cord.—In very rare instances diabetes mellitus or glycosuria is associated with symptoms indicating a gross lesion of the spinal cord, such as locomotor ataxia or disseminated sclerosis. In such cases the diabetes may be secondary to the nervous affection.

In another group of cases it is not clear whether the spinal disease is the cause of the diabetes, whether both are due to a common cause, or whether the association is merely accidental.

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Forms of Degeneration in the Posterior Columns of the Spinal Cord.

We have seen that pathological changes are found in the posterior columns in many affections. These lesions may be divided into four groups.

1. Changes secondary to lesion of the posterior nerve roots external to the cord (*see* p. 34).

2. Ascending degeneration following a complete or partial transverse lesion of the cord (*see* p. 35).

3. Degenerations of the posterior columns secondary to changes

in the fibres of posterior nerve roots occurring within the cord, and commencing directly these fibres have passed through the pia mater, at the point where they lose their neurilemma (or external sheath) and where also the myelin sheath is absent or greatly reduced for a very short interval. The degeneration is seen in the intra-medullary fibres of the posterior roots from the point where they have passed to the inner side of the pia mater. The changes affect the internal divisions of the intra-medullary posterior root fibres, whilst the external bundles of these fibres, which pass into Lissauer's tract, are usually not affected, or are affected only at a late period of the disease. Degenerated fibres of the posterior root may be traced into the posterior grey matter, and into Clarke's column. When the degeneration of the posterior root fibres is well marked in the lower part of the cord, degeneration is seen in the columns of Goll in the cervical region. (*See* Plate III).

Marked changes of this nature occur in tabes dorsalis and often in general paralysis of the insane. Similar but less extensive changes in the posterior root fibres, commencing at exactly the same point, are seen in many cases of intra-cranial tumour. I have also detected changes commencing at this point in several cases of diabetes mellitus (*see* p. 373) in a case of tumour of the spinal dura mater, and in one of alcoholic peripheral neuritis. Others have recorded these changes in cases of cachexia associated with cancer of various organs, after diphtheria, and chronic ergot poisoning.

In the peripheral nerves and spinal roots, lymph ascends to the cord in the inner meshes or lymph spaces of the fibrous perineural sheath. Toxins reach the cord by this channel, and pass for the most part along the nerve roots into the cord. In their extra-medullary portion these nerves are protected from the influence of the toxins by the vital action of the neurilemma sheath, but on losing this, in their intra-medullary part, they at once undergo degeneration (Orr and Rows).

4. In a fourth group the changes do *not* commence at the point where posterior root fibres pass through the pia mater, or at least they have not always been detected at this point. In this group are the changes seen in the posterior columns in combined postero-lateral degeneration (ataxic paraplegia, the sub-acute degeneration) in the spinal degeneration associated with severe anæmia, in Erb's syphilitic chronic spinal paralysis, pellagra, syringo-myelia, the peroneal type of muscular atrophy, and in "senile paraplegia." The slight changes occasionally found at the upper part of the cord in the columns of Goll in progressive muscular atrophy and amyotrophic lateral sclerosis, and the sclerotic patches of disseminated sclerosis sometimes found in the posterior columns, may be here mentioned.

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PLATE III.

INTRA-MEDULLARY FIBRES OF SPINAL POSTERIOR ROOTS IN SIX CASES. MARCHI'S STAIN.

Black dots = degenerated fibres.

FIG. I.—Degeneration of intra-medullary fibres of spinal posterior root in case of cerebellar tumour. Root just outside pia mater normal. Cervical region.

FIG. II.—Degeneration of intra-medullary fibres of posterior root. Tumour of third ventricle of brain. Cervical region.

FIG. III.—Degeneration of intra-medullary fibres of posterior root, early tabes. Lumbar region. Just outside pia mater root normal.

FIG. IV.—Degeneration of intra-medullary fibres of posterior root. Case of tumour of spinal dura mater.

FIG. V.—Degeneration of intra-medullary fibres of posterior root, diabetes mellitus: lumbar region. Just outside pia mater root normal.

FIG. VI.—Case of degeneration of posterior and lateral columns in severe anæmia. Posterior root fibres, intra- and extra-medullary, quite normal. To left of illustration are a few degenerated fibres in lateral column (black dots).

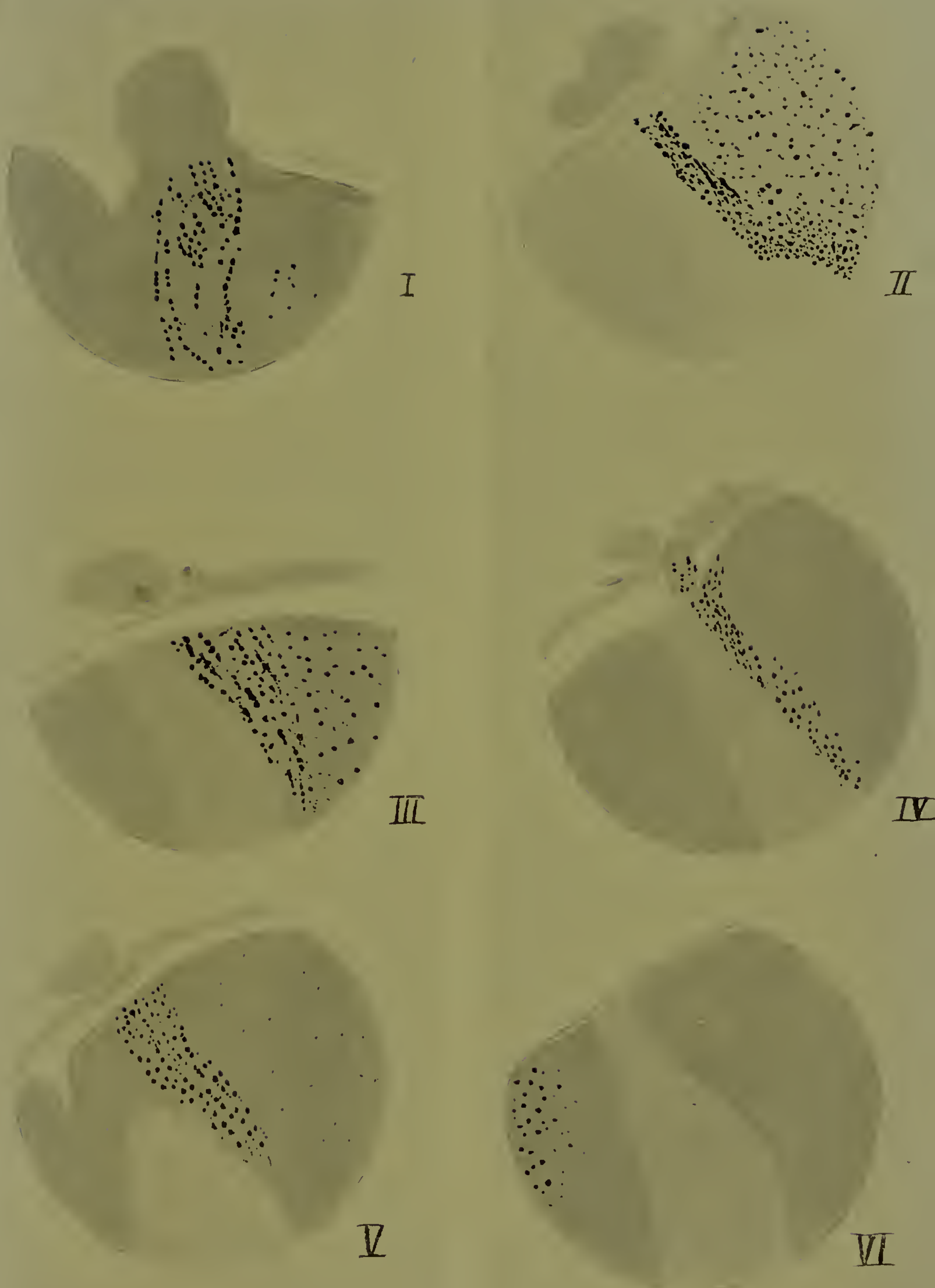

PLATE III.

SECTION XI

INFLAMMATION OF THE SPINAL MENINGES—SPINAL MENINGITIS

Inflammation of the dura mater is known as pachymeningitis (Greek : παχύς, thick ; μῆνιγξ, a membrane ; itis, suffix, to denote inflammation) ; inflammation of the delicate membranes as lepto-meningitis (Greek : λεπτός, thin ; μῆνιγξ, a membrane). In the latter condition the pia mater is inflamed, and usually the arachnoid is also affected. (Inflammation affecting the arachnoid, alone or chiefly, is known as arachnitis.)

Acute inflammation commencing in one membrane usually spreads to the others ; but *chronic* inflammation may be localised to one membrane.

External Meningitis.

(External spinal pachymeningitis, peri-pachymeningitis.)

This affection is an inflammation of the dura mater commencing on its outer side ; the inflammation does not usually extend to the inner side of the dura.

Etiology.—In most cases it is due to the extension of adjacent disease, purulent or non-purulent, to the dura mater. Most frequently external meningitis (usually localised and chronic) is secondary to tubercular caries of the vertebræ ; in very rare cases it is secondary to syphilitic vertebral caries. Sometimes acute external meningitis is caused by a deep sacral bed-sore or by a collection of pus in the neighbourhood of the vertebral column.

Acute general external (purulent) meningitis may be a primary disease according to some writers. Streptococci have been found in the purulent form of inflammation.

Pathology.—In the simple form, the outer side of the dura mater is red and opaque ; it is covered with pus when the inflammation is purulent. In chronic cases thick fibroid tissue and nodular thickenings may develop at the inflamed area. The fat external to the dura mater becomes absorbed, and the membrane often becomes adherent to the vertebræ.

When secondary to caries, the inflammation is usually localised : it may be purulent or caseous. When the affection is purulent and acute pus may extend all over the dura mater.

Symptoms.—Pain in the back is the chief symptom ; it is often in the lumbar region ; it is increased by movement ; and frequently there are

shooting or radiating pains in the limbs or around the trunk. Usually there is also spasm and rigidity of the muscles of the back. Other symptoms are hyperæsthesia of the skin, followed by anæsthesia and paralysis of muscles, from affection of nerve roots. Retention and, later, incontinence of urine may occur owing to paralysis of the bladder. When the affection is acute, there are general symptoms, such as rigors and high temperature. If much pus is formed there may be ascending paralysis, with loss of reflexes, anæsthesia and flaccid condition of the limbs. Bed-sores may develop later.

In the chronic localised form, pain is the chief symptom, and this is associated with symptoms of the primary disease and with those of secondary involvement of the cord.

Diagnosis.—The most important symptoms are those due to irritation of nerve roots, which may be followed by indications of compression of the cord. Acute external meningitis can rarely be distinguished from internal meningitis ; but a sign in favour of spinal meningitis being external would be deep œdema of the muscles of the back.

The probability of symptoms such as those just described being due to external meningitis would be increased by their association with certain pathological conditions—an abscess near the vertebral column, cellulitis, angina Ludovici, a bed-sore, and vertebral caries or necrosis.

Treatment.—If a localised collection of pus can be detected in the neighbourhood of the spinal canal, it should be opened surgically. Rest is important. Counter irritation is advisable in the chronic cases. The acute forms should be treated in the same manner as acute internal meningitis.

Tubercular external pachymeningitis is described on p. 143 and syphilitic meningitis on p. 389.

Internal Meningitis.

Meningitis commencing on the inner side of the dura mater is known as internal meningitis. The most important forms are :—

1. Acute leptomeningitis.
2. Chronic meningitis.
3. Hypertrophic cervical pachymeningitis.
4. Chronic syphilitic meningitis (pachymeningitis, leptomeningitis).

Internal meningitis may be limited to the pia mater and arachnoid (leptomeningitis) ; but in other cases it extends from these membranes to the inner side of the dura mater, or to the surface of the cord (meningo-myelitis). Internal or leptomeningitis may be acute or chronic.

Acute Spinal Leptomeningitis.

This affection is very rarely primary and simple. It may be traumatic, following fracture or dislocation of the vertebræ or other injuries. In some cases it is tubercular. On the Continent and in America epidemic cerebro-spinal meningitis is a comparatively common disease ; in Eng-

land this form is exceedingly rare, but cases of sporadic cerebro-spinal meningitis are occasionally met with resembling the epidemic form.

In most cases leptomeningitis is septic in origin. The septic form has followed septic infection from vertebral caries, from wounds, cellulitis, localised accumulations of pus, septic ulcers near the vertebral column, and septic processes in the uterus and pelvis. Also it has occasionally followed the acute infectious diseases, ulcerative endocarditis gonorrhœa and erysipelas. In rare cases it has followed septic middle ear disease, when the cerebral meninges have been almost unaffected. In most cases acute spinal leptomeningitis is associated with a cerebral affection of the same nature, but in other cases the spinal meninges are affected chiefly or exclusively.

Pathological Anatomy.—In the early stage the membranes are congested, and may be studded with points of ecchymosis. Later the pia mater and arachnoid are thickened and opaque from fibrinous or purulent exudations. These membranes become opaque, and grey or yellow in colour.

Microscopical examination shows that the pia and arachnoid are infiltrated with leucocytes, large round cells and spindle cells. The meningeal vessels are dilated and surrounded by round cells (*see* Fig. 172). The nerve roots are covered with cellular exudation. Usually the changes are most marked on the posterior surface.

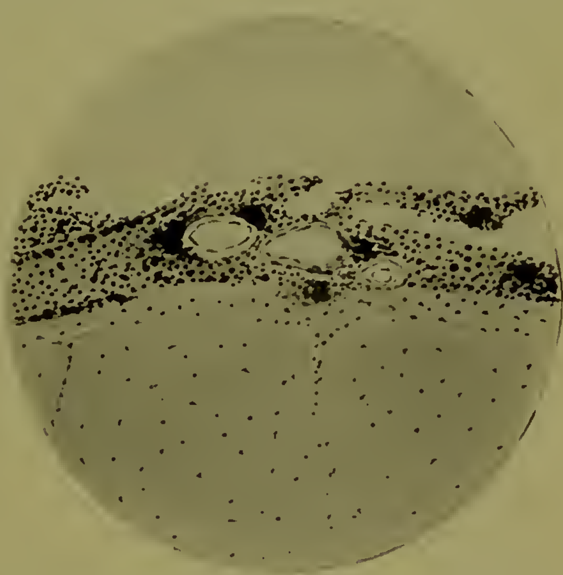

Fig. 172.—Posterior Surface of Spinal Cord with Pia Mater. Leptomeningitis (logwood stain). Note infiltration of pia mater with cells.

The surface of the cord may be affected from extension of the meningeal inflammation, and myelitic changes are then detected. (Marginal myelitis.)

In the tubercular form the membranes are studded with tubercles; the exudation is scanty and gelatinous, but not purulent. The inner side of the dura mater may also be studded with tubercles (*see* p. 386).

Septic or tubercular meningitis often extends from the brain to the cord.

Septic processes (associated with phlegmon, erysipelas, bed-sore, etc.) can extend from parts in the neighbourhood of the vertebral canal along the spinal vessels and nerves to the pia and arachnoid. The injection of streptococci into the sciatic nerve in rabbits has been followed by the extension of these organisms along the lymphatics of the nerves to the pia and arachnoid and spinal cord.

In rare cases of septic disease, as in malignant endocarditis, septic

emboli are conveyed to the pia and arachnoid, and septic meningitis extends from the meningeal vessels obstructed thereby.

Symptoms.—In many cases both spinal and cerebral meninges are affected, and the more prominent cerebral symptoms obscure the spinal symptoms. But in some cases the affection is spinal only, or chiefly spinal.

The symptoms commence acutely with shivering or rigors, pyrexia, severe pain in the back, and symptoms of irritation of sensory nerves. The pain varies in locality; it may be present in all parts of the back; it is more or less constant with exacerbations; it is increased by movement of the trunk; it radiates along the sensory nerves into the limbs and around the trunk as a girdle pain. Pressure on the spine and the application of a hot sponge to the skin increase the pain.

Irritation of motor nerves causes muscular spasms. The back muscles become rigid, the head and neck are often retracted, and opisthotonus sometimes occurs; the muscles of the abdomen, chest and limbs also become rigid.

FIG. 173.—Kernig's Sign in Meningitis. Leg cannot be extended on the thigh—cannot be moved upwards in direction of the arrow — when the patient is in the horizontal position as shown in diagram.

The skin becomes hyperæsthetic. The superficial and deep reflex are increased—especially the abdominal reflexes.

Kernig's sign can be detected, i.e. the patient is unable to extend the leg at the knee in the sitting position, on account of contraction of the flexors of the knee. This sign may also be detected in the following manner:—when the patient is lying in the horizontal position, if the thigh be flexed at the hip to a right angle with the trunk (i.e. brought into a vertical position) the leg cannot be extended fully on the thigh, owing to spasm and contraction of the muscles which flex at the knee joint.

Constipation and retention of urine are common. There is often dyspnœa; the pulse may be rapid or slow.

As the disease progresses paralytic symptoms develop. The limbs become weak or paralysed; sensation becomes diminished or lost; the bladder becomes paralysed, and the tendon reflexes disappear. Bedsores may develop.

The course is rapid and the prognosis very unfavourable. Death occurs in a few days in severe cases; in other cases within two or three weeks. Recovery occurs in rare cases.

Lumbar puncture may be of diagnostic value in doubtful cases. The puncture fluid in purulent meningitis is usually turbid, but not invariably so. The cells in the puncture fluid are increased, being usually polynuclear leucocytes in acute non-tubercular meningitis, but mononuclear lymphocytes in tubercular meningitis. The presence of

tubercle bacilli is clear evidence of tubercular meningitis, but the absence of tubercle bacilli is not conclusive that the affection is not tubercular (*see* p. 92). The presence of the diplococcus intra-cellularis points to epidemic cerebro-spinal meningitis. The presence of streptococci is in favour of purulent meningitis of other forms.

Diagnosis.—The symptoms of greatest diagnostic value are—pain in the back, rigidity of the muscles of the spine and neck, spasm of the muscles of the limbs, Kernig's sign, hyperæsthesia, the results of lumbar puncture, and the acute onset of the symptoms with pyrexia. The differential diagnosis is usually easy.

In *myelitis* pain in the back is slight or absent, and paralysis is usually an early and marked symptom.

Meningeal hæmorrhage is distinguished by the very sudden onset of the symptoms without pyrexia.

In *tetanus* there is no pyrexia at the onset, trismus is an early and marked symptom, and sensation is not affected.

Acute rheumatism may cause pain in the back, if the vertebral muscles and joints are affected, and thus raise the suspicion of meningitis. But in the latter disease swelling of the joints does not occur at an early period; also shooting pains in the limbs, affection of sensation, and bladder symptoms are in favour of meningitis.

Treatment.—Complete rest is necessary. The room should be kept quiet, and the light should be subdued. Bodily movements should be avoided as much as possible. The patient should lie upon the side rather than on the back. A water bed is desirable from the first. It is important to carefully attend to the feeding of the patient. Diaphoretic treatment (hot air or vapour baths), dry cupping to the spine, or the application of an ice bag to the spine have been recommended by various writers. Saline purgatives are probably of some service. Mercury is recommended by Sir William Gowers as the only internal remedy capable of influencing the disease: he advises that it should be given until the gums are slightly affected. "The oleate of mercury may be rubbed in along the spine." For the pain sedatives are required. Morphia (hypodermically in small doses $\frac{1}{12}$ to $\frac{1}{8}$ of a grain), codeia, chloroform inhalations, phenacetin, sulphonal, trional, belladonna and atropine (hypodermically) are of service. Atropin and morphia may be given together hypodermically. Chloral and bromides are of service also as sedatives. If the chloral is vomited back it may be given in an enema. When the disease is subsiding counter irritation to the spine may be employed. Paralytic conditions and other symptoms may be treated as in cases of myelitis.

Epidemic cerebro-spinal meningitis is not a strictly spinal affection, and will not be considered here. Excellent accounts of the disease are given by Leyden and Goldscheider (*Die Erkrankungen des Rückenmarks*. Wien, 1897) and by Osler in the Cavendish Lecture (*British Med. Journal*, 1899).

Chronic Spinal Meningitis.

Simple chronic spinal meningitis (excluding syphilitic meningitis and cervical pachymeningitis) is not a disease of much clinical importance. Usually it is found unexpectedly at the post-mortem examination on patients who have died of spinal or other diseases. It may be found in cases of tabes, myelitis, chronic alcoholism, and in senile individuals. Occasionally acute and epidemic meningitis have been followed by chronic meningitis. Localised thickening of the meninges is often found in caries of the vertebræ and in traumatic affections of the spine.

The chief symptoms of chronic meningitis are pain in the back, radiating pains in the limbs, rigidity of the spine, pain on movement of the back, paresis of muscles followed by wasting, and hyperæsthesia of the skin. There is no pyrexia. The symptoms of chronic lepto and pachy meningitis are so similar that a differential diagnosis between the two is scarcely possible.

Internal Hæmorrhagic Spinal Pachymeningitis.

The symptoms of this affection are generally complicated by those of a similar cerebral affection: usually they are those of a subacute cerebro-spinal meningitis. The chief symptoms due to the spinal affection are pain in the back, rigidity of the back, shooting pains and spasms in the limbs, and hyperæsthesia of the skin.

Often the patients are suffering from general paralysis of the insane, from chronic alcoholism or tubercular meningitis; sometimes the condition follows an injury.

Two forms of chronic meningitis are of much clinical and pathological interest, and require special consideration :—

1. Cervical hypertrophic pachymeningitis, and
2. Syphilitic meningitis (*see* p. 388).

Hypertrophic Cervical Pachymeningitis.

This form of meningitis was carefully described by Charcot and Joffroy many years ago. It is due to a chronic inflammation of the dura mater, which leads to the formation of a thick layer of new fibrous tissue on its internal surface. The affected dura mater becomes greatly thickened (from five to ten times its normal thickness). Often adhesions form between the dura mater and periosteum of the vertebral canal, and between the dura mater and the spinal cord. The new tissue also compresses the nerve roots and becomes adherent to them. The cord is finally affected and compression myelitis develops at the region of the thickened dura. At first only the surface of the cord may be inflamed (marginal or peripheral myelitis); later the whole transverse area of the cord may become sclerosed at the region of compression.

The disease occurs most frequently at the lower cervical region, although it may occur at other parts.

Etiology.—The disease has been attributed to cold, and to injury or strains. In some cases it is syphilitic in origin.

The **symptoms** of cervical pachymeningitis may be divided into three stages.

At first they are those of *meningeal inflammation and irritation* of the spinal nerve roots. The most prominent symptom is pain in the neck and between the shoulders. The pain radiates into the arms, and up to the back of the head. It is constant, with exacerbation or paroxysms of more intense pain. The neck is stiff, and there is pain on percussion of the cervical vertebral spines. Often there is paræsthesia and neuralgic pain in the distribution of the ulnar and median nerves. Tremor and slight spasm of the muscles of the arms is common.

After these symptoms have been present for two or three months *paralytic symptoms, of nerve root distribution,* gradually develop, and the pain subsides. The muscles supplied by the ulnar and median nerves become weak and gradually waste. Finally there is a well marked atrophic paralysis of these muscles.

The pain and paræsthesia in the arms is followed by diminished sensibility and finally by anæsthesia.

The small muscles of the hand and the flexors of the fingers and wrist usually become paralysed, whilst the extensors are spared. Hence the hands become hyper-extended at the wrist, and the fingers hyper-extended at the metacarpo-phalangeal joints but flexed at the phalangeal joints (combination of " claw-hand " and " preacher's hand ").

In the third stage the *spinal cord* becomes affected. The legs become weak and spastic and present the signs of spastic paraplegia or paraparesis. Sensation in the legs and trunk becomes diminished or lost, and bladder symptoms develop. The pains in the neck and arms subside, but the other symptoms may continue for years. Occasionally recovery occurs, but usually the spinal symptoms persist.

Diagnosis.—Pachymeningitis is distinguished from vertebral caries by the bony deformity and indications of bone disease, by the pain on movement, and by signs of tuberculosis in other parts of the body in the latter affection (*see* p. 150). The X-ray photograph may show a distinct deformity of diagnostic value in caries.

In cervical myelitis, amyotrophic lateral sclerosis and syringomyelia there may be spastic paralysis of the legs, with atrophic paralysis of the arms, as in cervical pachymeningitis ; but in the three affections first mentioned severe pains in the neck and radiating pains in the arms do not occur.

Treatment.—Mercury and iodide of potassium should be given when the affection is thought to be syphilitic in origin (*see* p. 399). Warm baths and salicylates have apparently been of service in some cases. Local treatment—painting the neck with iodine, or counter-irritation by the use of the actual cautery—has been recommended by many physicians. Drugs for the relief of pain (opium, morphia, antipyrin, pyramidon, etc.), may be necessary.

Spinal Tubercular Meningitis.

This affection is usually combined with the cerebral form. But the spinal symptoms are generally slight in comparison with the cerebral.

Pathological Anatomy.—In tubercular meningitis, tubercular disease is almost always present in some other part of the body, in the lungs, pleura, lymphatic glands, bones or joints, etc. In spinal tubercular meningitis there are two chief pathological changes—exudation and formation of tubercles. The meningitis is usually best marked at the upper part of the cord, and on the posterior surface. The pia mater and arachnoid are hyperæmic, turbid and thickened, and infiltrated with round cells ; but the exudation is not purulent. The walls of the blood vessels are thickened and may contain tubercles ; the adventitial coat is infiltrated with round cells. The pia mater is studded with miliary or sub-miliary tubercles. The dura mater is normal or only slightly affected. The nerve roots, especially the posterior, are infiltrated with cells.

The cord may present diffuse and irregular changes. Cell infiltration follows the septa and vessels from the periphery. Degeneration of nerve fibres and softening often occurs on the periphery of the cord (peri-myelitis, peripheral myelitis, or "rand-myelitis" of German writers). Secondary ascending and descending changes occur in the long tracts of the spinal cord. In many cases the pathological condition is therefore a tubercular meningo-myelitis.

The spinal **symptoms** correspond to those of acute or chronic spinal meningitis already described, but often myelitic symptoms are also present.

Diagnosis.—When the spinal symptoms are prominent in cerebro-spinal tubercular meningitis, or when spinal symptoms occur alone, the diagnosis is the same as that of the non-tubercular spinal meningitis. The tubercular nature of the affection is often indicated by signs of tuberculosis in other organs (lungs, pleura, etc.). In other cases these signs are absent, but the wasted cachectic appearance of the patient, persistent diarrhœa, enlarged glands, etc., would be in favour of the tubercular nature of the illness. Also the detection of tubercles in the choroid, with the ophthalmoscope, would be of diagnostic importance, though these are seen only in a few cases.

Another diagnostic sign is the presence of tubercle bacilli and lymphocytosis in the fluid obtained by lumbar puncture (*see* p. 93).

The disease is almost invariably fatal. The palliative treatment indicated in other forms of meningitis would be suitable.

Tuberculosis of the Spinal Cord.

In some cases the cell infiltration of the periphery of the cord, already mentioned, in tubercular meningitis is associated with tubercular infiltration.

In rare cases miliary and sub-miliary tubercles are found in the

cord substance without any tubercular meningitis. In some of these cases, in addition to the tubercles, there is a diffuse cell infiltration, along with patches of softening and hæmorrhage and extensive areas of degeneration—the condition being practically a tubercular myelitis. In other cases the diffuse inflammatory changes are absent, and the condition is simply a disseminated tuberculosis of the cord.

A special form of spinal tuberculosis is the solitary tubercle or conglomeration tubercle, which produces the symptoms of spinal tumour. The caseous mass is formed by the union of a number of miliary tubercles; its size varies considerably—occasionally it is as large as a cherry. The cord, around the tubercular mass, presents changes similar to those around a spinal tumour (*see* p. 163).

* * * * *

The following are the affections of the spinal cord directly or indirectly due to tubercular disease, given in tabular form :—

TUBERCULOUS DISEASES OF THE SPINAL CORD AND MEMBRANES.

1. Caseous external pachymeningitis, associated with vertebral caries or tubercular tumour growing into the vertebral canal.
2. Tubercular tumour of the dura mater (extra- or intra-dural). Vertebral column unaffected.
3. Tubercular tumour of the pia or arachnoid, or between the meninges.
4. Tubercular spinal meningitis.
5. Tubercular meningo-myelitis.
6. Tubercular meningitis with tubercular infiltration of the cord.
7. Miliary tuberculosis of the cord—(*a*) with cell infiltration and softening (tubercular myelitis) ; (*b*) without inflammatory changes—disseminated tuberculosis of the cord.
8. Solitary or conglomerate tubercle.

The most common affection of the cord caused indirectly through tuberculosis is compression myelitis secondary to vertebral caries ; the compression being due to various pathological conditions—external pachymeningitis, tubercular abscess, or very rarely to displaced bone ; in rare cases there is tubercular infiltration of the cord (*see* p. 143). The most frequent form of tubercular disease of the cord itself is (8) the solitary or conglomerate tubercle. (Symptoms mentioned on p. 169.)

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Schlesigner. Article, "Tumoren des Rückenmarks und seiner Hüllen." *Handbuch der pathologischen Anatomie des Nervensystems.* (Flatau, Jacobsohn, and Minor) Abth iv., Berlin, 1903. (Tubercular diseases of the cord.)

Meningeal hæmorrhage is described on p. 191, and meningeal tumour on p. 159.

SECTION XII

THE CLINICAL FORMS AND PATHOLOGICAL ANATOMY OF SPINAL SYPHILIS

Acquired syphilis produces various forms of spinal disease, according to the distribution and nature of the pathological lesions. These forms of spinal syphilis often closely resemble affections of a non-specific nature, but their recognition is of much practical importance, especially with reference to treatment; and the varied pathological lesions with which they are associated are of considerable interest.

I. **Symptoms of compression of the spinal cord or nerve roots may be produced by syphilitic disease of the vertebræ (syphilitic caries, necrosis, gumma, periostitis, or osteitis).**—This is an exceedingly rare form of spinal syphilis, and may at first be mistaken for tuberculous caries or tumour of the vertebræ. The symptoms resemble those of "compression myelitis" from other causes. The diagnosis is based on—(1) the evidence of disease of the vertebræ, with or without implication of the nerve roots and spinal cord; (2) the absence of indications of tuberculous disease, cancer, or tumour growth in any part of the body; (3) the evidence of previous syphilitic infection; (4) the good results of antisyphilitic treatment.

II. **Meningitis (pachymeningitis and leptomeningitis).**—In another very rare form there are symptoms of *chronic meningitis*, without indications of involvement of the spinal cord. Pathologically, chronic pachymeningitis and leptomeningitis have been found; and generally the altered dura mater, arachnoid, and pia mater have been fused together so as to form one thick sheath of adherent fibrous or gummatous membrane around the cord. Often the changes are limited to, or most pronounced at, one region of the cord, as in the lumbar or cervical region. Hence the symptoms are sometimes most marked in the legs, sometimes in the arms. Pain in the back is the most important symptom. It is often worse at night (like syphilitic headache). The pains radiate into the limbs, and other symptoms of involvement of nerve roots develop. The temperature is normal or only very slightly raised (to 100° F.). The evidences of previous syphilitic infection, the absence of fever, the absence of any indication of tuberculosis or of spinal caries, are important points in the diagnosis.

When the meningitis is in the cervical region the symptoms resemble those of cervical hypertrophic pachymeningitis (*see* p. 385). At a very

late stage spastic paresis of the legs may develop if the cord is compressed or implicated. When the meningitis is in the lumbar region the pain is in the legs chiefly, and at a late period paresis may develop.

When the meningitis has involved only the nerve roots of the cauda equina the symptoms have been chiefly pain radiating in the distribution of the sacral plexus, and in the region of the bladder and genital organs, and afterwards anæsthesia of these parts, with paralysis of the bladder and rectum and loss of sexual power.

III. **Meningomyelitis.**—This is the most common form of spinal syphilis.

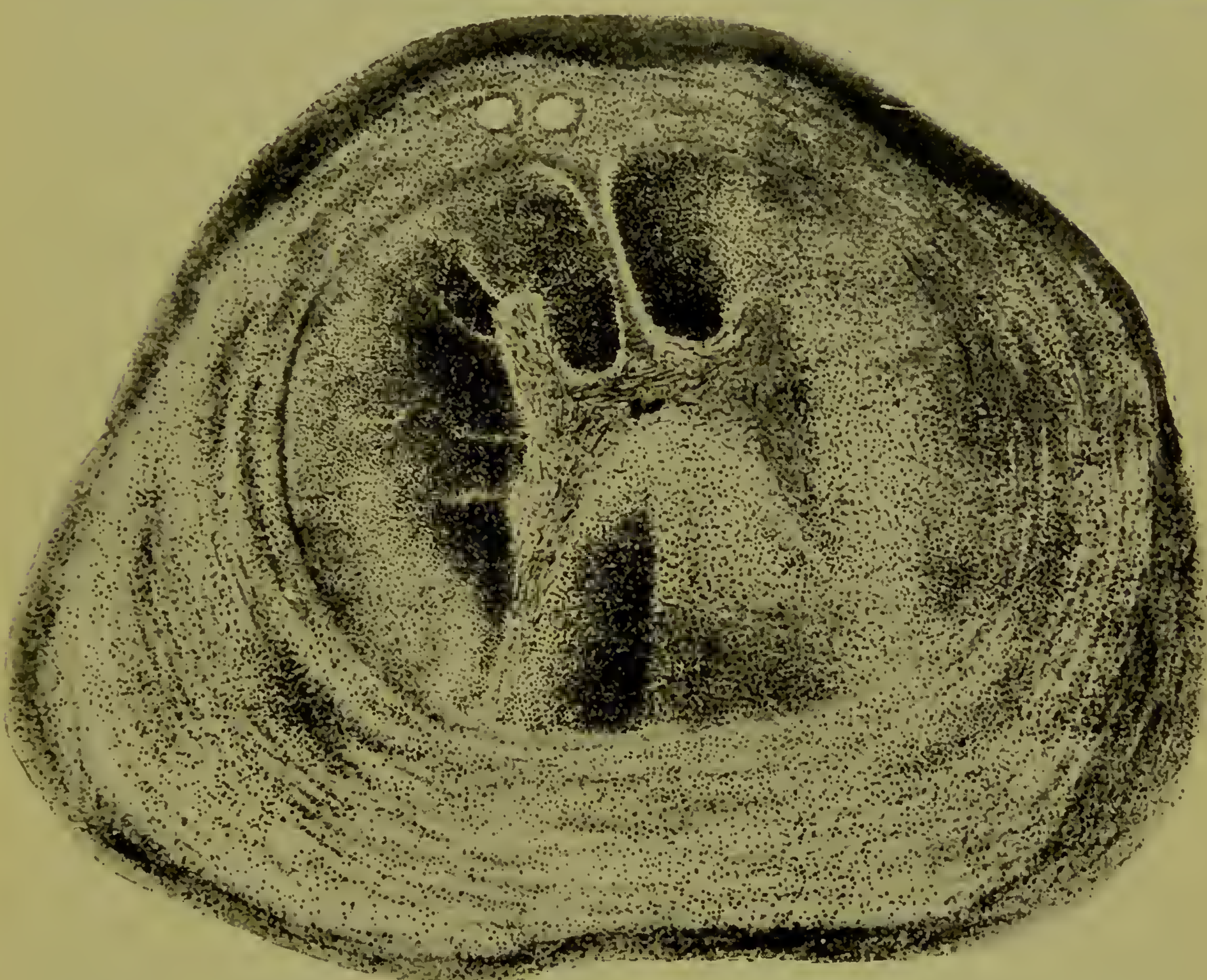
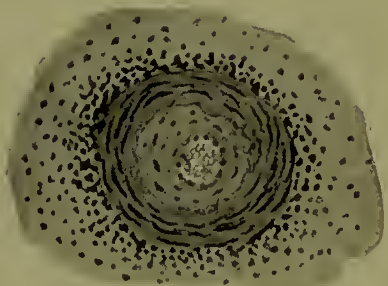


FIG. 174.—Syphilitic Meningitis, dorsal region. Pachymeningitis and leptomeningitis. All membranes united to form a fibrous sheath around the cord (after Homén, *Archiv. f. Dermat. und Syph.* Wien. Bd. xlvi. 1898).

There are meningeal symptoms first (pain in the back, and indications of implication of spinal nerve roots), followed in course of time (weeks or months) by symptoms of involvement of the spinal cord. The pain may radiate into the legs or into the arms or around the trunk (girdle pains), according to the chief seat of the disease. By implication of nerve roots localised paresis and anæsthesia are sometimes produced. The cord symptoms consist of paraparesis or paraplegia, usually with rigidity, increase of the deep reflexes, ankle-clonus and the extensor (Babinski) type of plantar reflex, and frequently with bladder symptoms and sensory

disturbances. There may be partial or complete anæsthesia to all forms of sensation; but often some forms are affected and others spared; sometimes *loss of sensation to temperature* (*especially to cold*) is the chief or only sensory disturbance. In a number of cases I have found the sensation for temperature affected before other forms of sensation. Hence it is important to test the temperature sense (especially sensation for cold) in all cases of suspected spinal syphilis, before deciding that sensation is normal. The extent of the sensory affections will vary with the level of the lesion. In a case examined recently, I found loss of the vibrating sensation the only form of anæsthesia; in another case the vibrating sensation was lost over a much greater area than other forms of sensation. Bladder symptoms may occur before the paresis of the legs. In some cases bed sores and other complications of myelitis develop finally.

In the diagnosis of syphilitic meningomyelitis the points in favour



FIGS. 175 and 176.—Small Meningeal Arteries from a case of Syphilitic Meningo-myelitis. Endarteritis and periarteritis. Logwood and eosin.

of a specific affection mentioned on p. 398 are important. The history of pain in the back for a considerable time before the paraplegia, and the absence of signs of vertebral caries, are valuable indications in the diagnosis from acute primary myelitis and compression myelitis.

Pathologically, both the meninges and the cord are affected. Syphilitic disease of the blood vessels usually plays an important part in the pathological anatomy. The spinal arteries may present syphilitic endarteritis or periarteritis, or both may be combined. The veins are affected in a similar manner (endo- and periphlebitis). Thrombosis is occasionally observed in the spinal veins or arteries, and endophlebitis obliterans of the meningeal veins is sometimes present (*see* Figs. 175 and 176). Miliary gummata may be seen in the walls of the spinal veins, and occasionally in the walls of the arteries. The vascular changes occur both in the meninges and within the spinal cord. The pia mater and arachnoid present cell infiltration chiefly around the vessels; in some cases there is a gummatous infiltration of the membranes, and in other cases a fibrous thickening. The nervous elements of the spinal cord are damaged (1) by cell infiltration or gummatous infiltration, (2) by softening or degeneration, following partial or complete obliteration of the blood vessels, owing to thickening of the vessel walls or to thrombosis.

The changes in the nervous elements are often limited to the periphery of the cord, or they may extend over a large portion of the transverse section; but the peripheral white matter is usually most affected, and the grey matter is frequently spared. From the inflamed pia mater broad septa of cell infiltration, or fibrous tissue, or wedge-shaped masses of gummatous infiltration, often invade the periphery of the cord (*see* Fig. 177). The nerve cells and fibres break down and degenerate in the affected areas, just as in non-specific myelitis. Also, these regions are often infiltrated with leucocytes and compound granular cells, and frequently the perivascular spaces are distended with round cells. All the changes are usually most marked in the dorsal region of the cord, and though irregular in their exact distribution, they affect chiefly the part supplied by the peripheral arteries of the cord (*see* Fig. 26). Secondary ascending and descending degeneration follows the spinal softening described.

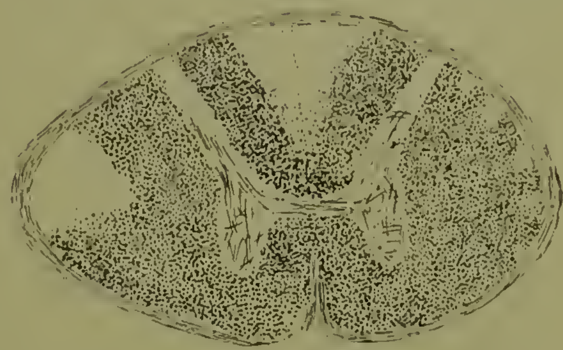


Fig. 177.—Syphilitic Meningo-myelitis. Weigert's stain. Wedge-shaped mass of gummatous infiltration in the lateral column (left-hand side). Meninges thickened.

IV. **Acute syphilitic paraplegia (acute syphilitic myelitis).**—Sometimes the symptoms of spinal syphilis resemble those of acute transverse myelitis, or even spinal hæmorrhage. There is a sudden onset of paralysis of both legs, with paralysis of the bladder and rectum. The intercostals may also be paralysed; occasionally, but rarely, the paralysis extends to the arms. There may be loss of all forms of sensation or analgesia and thermo-anæsthesia only; or thermo-anæsthesia, whilst other forms of sensation are normal. Frequently the sensory disturbances are slight, whilst the motor are very well marked. The knee-jerks may be present or absent. Ankle-clonus and spastic symptoms may occur, but in the cases which have come under my own observation ankle-clonus has usually been absent. Prodromal symptoms such as pains in the back or limbs, girdle sensation, etc., are often present for a short time. Prodromal symptoms are noticed for a longer period before the onset of paralysis in syphilitic cases than they are (when present) in non-specific myelitis. Retention of urine may occur for some days before the development of paraplegia. Though suggestive, this point is not quite diagnostic. Often cases of acute paraplegia occur at a comparatively early date after the syphilitic in-

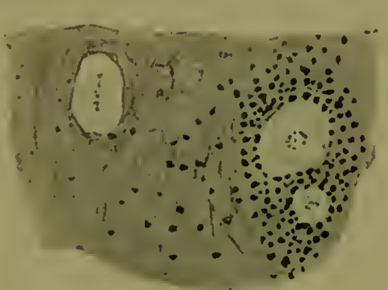


Fig. 178.—Hyaline Vessels from case of Syphilitic Meningo-myelitis. Vessels to the right surrounded by cell infiltration; that to the left by sclerosis.

fection. I have seen one case seven months and another case twelve months after infection.

The records of pathological examination in these cases are not numerous. In one which I examined a few years ago the chief change was marked disease of the blood vessels of the meninges and of the cord—endarteritis, periarteritis, endophlebitis and periphlebitis. Numerous vessels in the grey matter and adjacent white matter of the cord were greatly dilated and obstructed by thrombi which were undergoing organisation. Around the thrombosed vessels were hæmorrhagic infiltration, softening (degeneration and breaking down of nerve elements), and round-celled infiltration. There was also slight leptomeningitis. All the changes were most marked in the middle dorsal region and they resembled pathologically those produced by syphilitic cerebral thrombosis. Evidently the paraplegia had been due to thrombosis in blood vessels, within the spinal cord, which were affected with syphilitic disease (*see* Plates IV, V and VI). The thrombosis had been followed by spinal hæmorrhage and softening. In a number of other cases on record, the paraplegia has been due to spinal softening, following marked syphilitic disease of the spinal blood vessels, with great thickening of the vessel walls, but without thrombosis. In a third group of cases the pathological changes have been the same as in non-specific myelitis, and the spinal blood vessels have presented no sign of syphilitic disease. Possibly in this third group of cases, the lesion has sometimes been an ordinary non-specific myelitis, occurring in an individual who has happened to have suffered previously from syphilis.

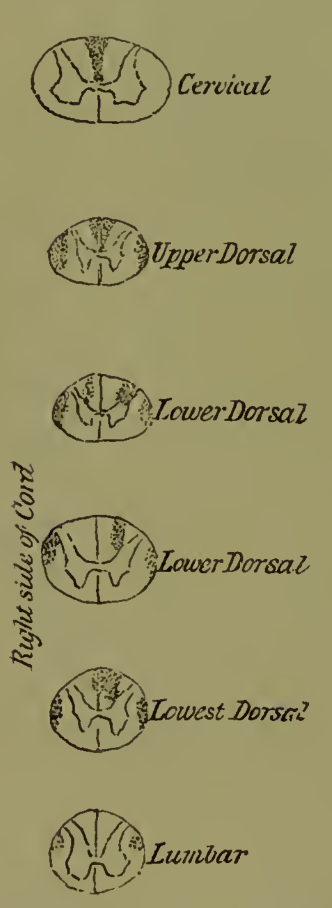


FIG. 179.—Showing distribution of lesions in the Spinal Cord at various levels. The sclerotic and degenerated parts are shaded. Erb's syphilitic spinal paralysis.

V. **Erb's syphilitic spinal paralysis.**—In 1892, Erb drew attention to a class of cases of chronic spinal syphilis, presenting a certain group of symptoms; he believes that they form a common and distinct clinical variety of spinal syphilis. The patients present the familiar symptoms of spastic paresis or paralysis, as regards gait, attitude and movements. The patellar tendon reflexes are increased, ankle-clonus is present, and there is the extensor (Babinski) type of plantar reflex, but there is relatively only slight muscular rigidity. The bladder is constantly affected (slight or marked incontinence or retention). As a rule there is only slight, though constant, subjective and objective affection of sensation :—diminution of the various forms of cutaneous sensibility to touch, pain and temperature. In some cases I have found loss of the

PLATE IV.

Sections of Spinal Cord, showing Position of Hæmorrhage (brownish-red) in Case of Acute Syphilitic Paraplegia.

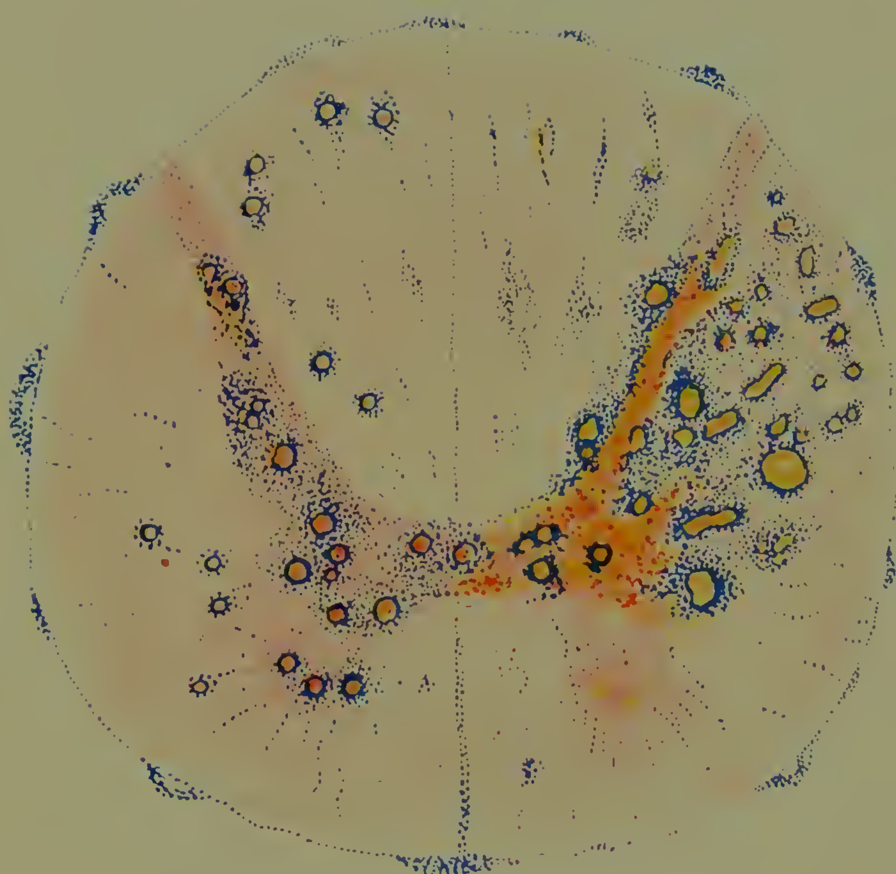


PLATE V.

Syphilitic Thrombosis and Hæmorrhage in the Spinal Cord. Case of Acute Syphilitic Paraplegia ("Myelitis"). Very Low Power of Microscope. Stained with Logwood and Eosin.

Section shows dilated and thrombosed blood-vessels, surrounded by cell infiltration (blue); hæmorrhage and blood within dilated vessels coloured red.

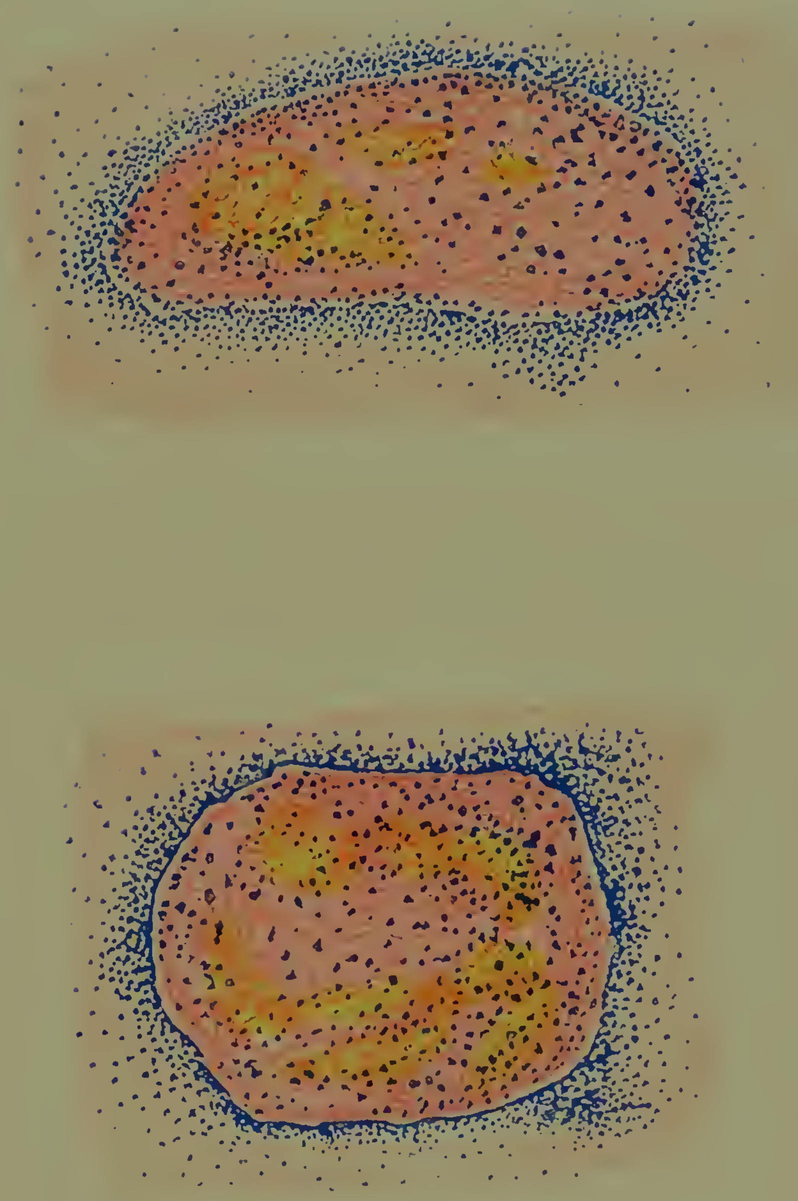


PLATE VI.

Sections of two Dilated and Thrombosed Blood-vessels of the Spinal Cord (represented in Plate V). Highly magnified.

Syphilitic thrombosis (syphilitic "myelitis"). Stained with logwood and eosin. Leucocytes and cell infiltration blue; organized clot within the vessels pink; blood yellow; cord substance around vessels pink.

sensation of temperature only, or loss of the vibrating sensation only, whilst other forms of sensation were normal (*see* p. 80). Pain is slight. The onset of the disease is gradual, seldom rapid. Marked paresis gradually develops, but only exceptionally is there complete paraplegia. The upper half of the body is unaffected.

FIG. 180.—Section of Spinal Cord in Upper Dorsal Region. Weigert's stain. Pale parts = regions of sclerosis; black dots = normal nerve fibres. Erb's syphilitic spinal paralysis.

This form of spinal syphilis is distinguished from primary lateral sclerosis by the presence of disturbances of sensation and of the bladder functions; from transverse myelitis by the absence of marked paralytic phenomena, bed-sores, and cystitis; and from meningo-myelitis by the absence of meningitic and root symptoms.

There has been considerable discussion as to whether "Erb's spinal syphilitic paralysis" should be regarded as a special disease of the spinal cord or not; but cases corresponding to his description form a clinical group to which the name just given may be conveniently applied.

The pathological anatomy of this group has not yet been definitely decided, owing to the few autopsies which have been recorded. In a case in which I made the pathological examination some years ago, there were syphilitic changes distributed somewhat irregularly in the cord and chiefly in the white matter. A most prominent change was endarteritis and hyaline degeneration of the arteries of the spinal cord, and meninges. There was slight meningitis. In the upper dorsal regions there was a large wedge-shaped patch of gummatous infiltration in the right antero-lateral column, extending from the surface of the cord into the grey matter. There were several irregular sclerotic patches with one patch of cell infiltration (gummatous), in the posterior columns in the lowest dorsal region. In the whole of the dorsal region there was sclerosis at the periphery of the cord just under the pia mater and this slightly invaded the lateral pyramidal tracts. There was sclerosis in the posterior median columns in the cervical and upper dorsal regions

FIG. 181.—Section of Spinal Cord in Lowest Dorsal Region (Weigert's stain); affected parts pale; normal nerve fibres represented by black dots and lines. Erb's syphilitic spinal paralysis.

and descending sclerosis in the lateral pyramidal tracts in the lumbar region (*see* Figs. 179, 180 and 181).

In a case of syphilitic paraplegia, which I recorded some years ago, there was combined degeneration of the posterior and lateral columns of the cord. During life the chief symptoms were paralysis of both legs, with rigidity, and bladder and rectal symptoms. The arms were not affected. The paralysis and rigidity of the legs were very well marked ; in this respect and in other points, the affection did not quite correspond clinically with Erb's group of cases. Death did not occur until nine years after the paraplegia developed. Pathological examination of the spinal cord showed degeneration (sclerosis) of the crossed pyramidal tracts throughout the *whole length of the cord*, with degeneration of Goll's columns in the cervical region (increasing in extent from below upwards), degeneration of the direct cerebellar tracts above the middle dorsal region, and degeneration of the periphery of the cord in the cervical region extending a little farther forwards than the region of the direct cerebellar tracts (*see* Plate VII).

In the few cases of Erb's spinal syphilitic paralysis in which a *post-mortem* examination has been obtained, a combined grey degeneration has been found chiefly in the posterior half of the lateral tracts—the pyramidal tracts, the direct cerebellar tracts and the tracts of Gowers ; also in the posterior tracts—the tracts of Goll and part of Burdach's columns.

Erb concludes that the pathological basis of the clinical type which he has described, is a combined system disease of the lateral and posterior tracts, either alone or accompanied by a somewhat diffuse, patchy, transverse lesion in the dorsal cord (as in the case I have just described, p. 393).

Nonne has analysed the cases recorded and concludes that the symptoms may be caused by several lesions :—

1. Chronic myelitis in patches, with ascending and descending degeneration.
2. These changes combined with a primary degeneration of the pyramidal tracts.
3. Degeneration of the pyramidal tracts alone.
4. A combined primary postero-lateral tract degeneration, affecting the posterior columns, the crossed pyramidal tracts, the direct cerebellar tracts and the tracts of Gowers.

VI. **Gumma of the spinal cord, or meninges.**—In rare cases of spinal syphilis the symptoms have been those of a localised meningeal or intramedullary spinal tumour, and a gumma has been diagnosed. In a comparatively few cases only, the diagnosis has been verified pathologically, and a gumma has been found, post-mortem, in the meninges, or more rarely within the cord. Often there have been also other syphilitic changes in the cord.

When the gumma has commenced in the meninges, the symptoms have been chiefly pain, with signs of irritation of nerve roots first, and

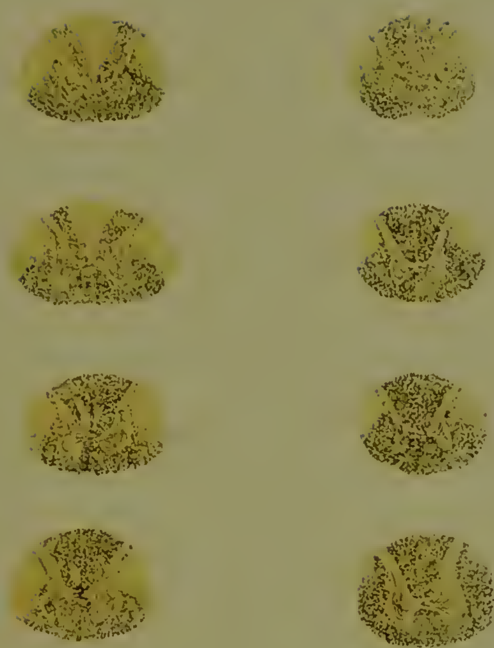


PLATE VII.

SYPHILITIC PARAPLEGIA. WEIGERT'S STAINS.

Sclerosis in posterior and lateral columns (sclerosed parts pale brown).

Figures arranged in the following order :—

Upper cervical regions.	Dorsal region.
Middle ,, ,,	Dorsal ,,
Lower ,, ,,	Dorsal ,,
Junction of cervical and dorsal.	Lumbar ,,

paraplegia at a later date. When the gumma has commenced within the spinal cord, the root symptoms have been usually absent, and clinically the case has presented the symptoms of a rather slowly developing paraplegia. Sometimes there have been symptoms of Brown-Séquard's paralysis (hemiparaplegia) at first in cases of gumma of the meninges or cord.

The diagnosis, pathologically, from a tubercular mass is not always easy. In favour of gumma are other syphilitic lesions or symptoms, the very thick, firm fibrous capsule of the nodule, the situation on the surface, or in the meninges, of the cord, and the absence of pus in the centre of the lesion. In favour of a tubercular mass would be the presence of tubercular disease in other parts of the body, the presence of small or miliary tubercles around the larger mass, the situation of the lesion near the centre of the cord, and the presence of pus in the centre of the mass. Tubercle bacilli if present would be diagnostic of tubercular lesion; their absence is not conclusive evidence.

FIG. 182.—Transverse Section of the Spinal Cord, showing the position of a gummatous tumour with surrounding myelitis.

VII. **Anomalous forms.**—In addition to the clinical forms already mentioned, there are a number of other rare varieties, in which the symptoms often resemble those produced by non-specific cord lesions :—(*a*) Cases of Brown-Séquard's paralysis (hemiparaplegia), due to a unilateral lesion of the cord such as a localised unilateral gummatous meningomyelitis. (*b*) Cases of triplegia, in which both legs and one arm are paralysed—hemiplegia along with paraplegia ; these are due to the combination of a unilateral cerebral lesion with a bilateral spinal lesion. (*c*) Cases somewhat simulating disseminated sclerosis, in the evidence of multiplicity of lesions ; these are due to multiple syphilitic lesions. But in the syphilitic cases true nystagmus, true intention tremor, and scanning

speech are absent, whilst meningeal symptoms and immobility of the pupils may be present. (*d*) Cases simulating lateral sclerosis; due to secondary sclerosis of the crossed pyramidal tracts of the cord, with slight changes elsewhere. (*e*) Cases simulating anterior poliomyelitis. Though the lesion has not been verified post-mortem, it is probable that there have been changes in the area of distribution of the central arteries of the spinal cord—anterior median arteries. (*f*) Cases simulating amyotrophic lateral sclerosis or progressive muscular atrophy; due to meningomyelitis in the lower cervical region. (*g*) Cases simulating pseudo-hypertrophic paralysis in the gait and manner of rising into the erect posture, but differing in the severe pain in the back and limbs, and in other symptoms; probably such cases are due to chronic meningitis in the lumbar and sacral region of the cord, producing paresis of muscles about the hips. (*h*) Cases simulating syringomyelia as regards the sensory symptoms: due to meningomyelitis of special distribution. There are also cases, presenting symptoms of meningomyelitis during life, which simulate syringomyelia on superficial examination pathologically. In such cases pathologically, there is meningomyelitis, with cavities (due to softening) in the grey matter of the cord. (*i*) Cases simulating locomotor ataxia (syphilitic pseudo-tabes); pathologically these are due to a meningomyelitis invading the posterior columns of the cord, or to a gummatous infiltration of the posterior columns.

In all these anomalous forms, the diagnosis is usually made readily, if the patient be carefully examined. Some symptoms are present in the specific cases which are always absent in the non-specific diseases, or some of the most characteristic symptoms of the non-specific disease are wanting.

VIII. Finally, there is the question of the **syphilitic origin of true locomotor ataxia.** Pathologically, the changes in this disease are not of a syphilitic nature in the strict sense, and the disease can only be regarded as a post-syphilitic degeneration. The relation of this disease to syphilis is discussed on p. 292.

IX. In children suffering from **hereditary syphilis** spinal symptoms are occasionally observed corresponding to those of meningomyelitis, Erb's spinal syphilis, pseudo-tabes and multiple lesions. Pathological examination reveals various lesions (gummata, endarteritis, myelitis, sclerotic changes). Frequently cerebral changes are also present, and very often the cerebral symptoms are much more marked than the spinal.

Congenital spastic paraplegia (Little's disease—diplegia) has been attributed to hereditary syphilis in some cases.

Juvenile tabes is undoubtedly due to hereditary syphilis (*see* p. 294), or to syphilitic infection at a very early period of life.

It has been shown also that various forms of infantile localised paralysis (monoplegia) may be due to spinal meningitis with involvement of the anterior nerve roots.

In all of these cases signs of hereditary syphilis (Hutchinson's teeth,

choroiditis, etc.) are important indications of the syphilitic nature of the spinal affections.

Frequency of the various forms of Spinal Syphilis.—In 35 cases of spinal syphilis which have come under my observation, the forms of disease were as follows :—

Syphilitic disease of the vertebræ	0
Chronic syphilitic meningitis	3
Meningomyelitis	16
Acute paraplegia (acute syphilitic myelitis)	6
Chronic syphilitic spinal paralysis (Erb's form)	6
Gumma of the cord (verified)	1
Triplegia	1
Brown-Séquard's paralysis	1
Pseudo-tabes	1
	35

The etiology and general pathology of spinal syphilis.—Spinal syphilis (excluding tabes) is a somewhat rare affection. During the ten years the writer held the post of Medical Registrar at the Manchester Royal Infirmary, there were 14,575 medical in-patients ; of these, 2,456 suffered from diseases of the nervous system ; 118 of the nervous cases suffered from tabes, but only 32 from spinal syphilis.

Males are much more frequently affected than females. In the cases to which I have referred, there were 26 males and 5 females. The age of the patient is most frequently between 20 and 40.

The spinal affection may occur a short time after syphilitic infection (seven months), or not for many years (ten or fifteen). In 17 out of 27 cases, the disease had occurred within the first five years after infection ; in 2 cases within the first twelve months.

Some neurologists believe that there is a special form of syphilis which is particularly liable to be followed by lesions of the central nervous system, and there is considerable evidence in favour of this view.

The pathological changes produced by syphilis may be divided into three classes—(1) Those which indicate syphilis most clearly—gummata, or gummatous infiltration of the meninges or cord ; minute gummatous nodules in the walls of the vessels ; and inflammation of the meninges, followed by caseous and fibroid changes. (2) Changes which are very suggestive, but not quite conclusive, of syphilis—disease of the blood-vessels, endarteritis, and periarteritis, endophlebitis and periphlebitis. (3) Changes secondary to the vascular disease ; thrombosis of spinal vessels, or complete or partial obstruction of the vessels, from thickening and disease of their walls. As a result of either of these two vascular changes, softening and degeneration of nerve elements occur ; occasionally spinal hæmorrhage is produced. Following the destruction of the nerve elements, ascending and descending sclerosis gradually develop.

The lesions produced by syphilis in the nervous system may be also divided into three other groups : (1) Those in the adventitial tissue—blood vessels, connective tissues, and membranes ; (2) degeneration

in the nervous elements (fibres and cells) secondary to the changes in first group; and (3) post-syphilitic degeneration of fibres and cells.[1]

In some of the cases of myelitis in syphilitic individuals, in which there are no evidences of syphilitic changes in the cord, or its vessels, and the microscopical appearances are the same as in non-syphilitic myelitis, it is a disputed point whether the lesions are to be attributed to syphilis, or whether they are to be regarded as non-specific lesions occurring in an individual who happens to have suffered from syphilis.

From my own pathological observations, I am inclined to attach the greatest importance to syphilitic vascular changes in the pathology of spinal syphilis.

The **diagnosis** of spinal syphilis is of great importance, especially at the early stage, when good results may be expected from prompt treatment. The diagnosis of each variety of spinal syphilis will require separate consideration, with reference to the non-specific affections it most closely simulates. The following general indications are, however, in favour of the syphilitic nature of a spinal disease :—

(1) The history of previous syphilitic infection. (2) Signs of present or previous syphilitic disease in various parts of the body :—cicatrix on the penis, circular scars of old ulcers on the legs or face, perforation of the soft palate, etc. (3) The Argyll-Robertson pupil, complete immobility, irregularity of the outline, or eccentrical position of the pupils (*see* p. 308) are very important indications in favour of past syphilis. Other valuable ocular signs are: Evidences of old iritis, choroiditis, choroido-retinitis, syphilitic retinitis, and fine opacities in the vitreous. (4) The presence of cerebral symptoms (due to associated syphilitic cerebral disease). (5) The relatively slight intensity of the cord disease as compared with the extensive area involved (Sachs). (6) The presence of Brown-Séquard's paralysis—hemiparaplegia—at some period of the illness. (Often this group of symptoms is incomplete and temporary.) (7) Fluctuations in the intensity of nervous symptoms. (8) Multiplicity of lesions. (9) Several French authors attach much importance to pain in the back which is worse at night. This symptom I have observed in a number of cases.

If the cerebro-spinal fluid, obtained by lumbar puncture, contains an excess of cells which have the character of lymphocytes (lymphocytosis), this feature would be in favour of the diagnosis of spinal or cerebral syphilitic disease, tabes, general paralysis of the insane, or tubercular disease of the central nervous system (*see* p. 93).

The improvement under antisyphilitic treatment may, in certain cases, be regarded as evidence in favour of the syphilitic nature of the disease.

The **prognosis** in spinal syphilis is, on the whole, better than in other affections of the spinal cord. But it differs according to the form of the

[1] *See* Lecture by Sir Wm. Gowers, *Brit. Med. Journal*, April 4, 1903.

disease. Complete or partial recovery occurs in some cases, but others terminate fatally. The prognosis is better when the meninges are chiefly affected, but the more the cord substance is involved, the less are the prospects of recovery. The prognosis is worst in the cases of acute paraplegia ("acute syphilitic myelitis"), especially when the bladder and rectum are paralysed. Some of these cases rapidly terminate fatally.

The causes of death are the same as in myelitis (*see* p. 135).

Sir William Gowers points out that syphilitic disease of the nervous system, "developing in what may be termed 'adventitial' elements, produces symptoms for the most part through the changes it causes in the nerve elements themselves. But these are simple, not specific. They are secondary to the syphilitic disease." Specific treatment acts only on the specific process. It has no direct influence on the secondary changes in the nerve elements, and little, if any, on the post-syphilitic degenerations. The possibility of recovery in case of slow compression depends partly on its degree, but still more on its duration. Paraplegia from such pressure may be complete for a month, and in time recovery may be perfect. If complete for three months recovery may possibly be perfect, but is more likely to fall short of the normal state ; but if complete paraplegia from compression has endured for six months only partial recovery can be expected ; and after a year the return of function may be very slight, but at best will not be great. The more rapid the development of the symptoms the less is their course affected by specific treatment (Gowers).

In the thirty-two cases already referred to death occurred in nine, recovery in ten ; the other cases passed from observation without any great change in their condition.

The nature of the nine fatal cases was as follows—acute paraplegia ("acute myelitis"), five cases ; Erb's chronic syphilitic paralysis, one case ; gumma of the cord, one case ; triplegia, one case ; meningo-myelitis, one case. The nature of the ten cases which recovered was as follows—five of meningo-myelitis, three of meningitis, one of acute paraplegia, one of pseudo-tabes.

Treatment.—At the early stage of spinal syphilis, especially when the meninges only are involved, treatment is of the greatest importance. It is then that we may hope to arrest the syphilitic changes. When the cord has been seriously damaged, and nerve elements destroyed and replaced by cicatrical tissue, we cannot hope to remove this scar tissue by treatment, or to cause new nerve fibres or cells to develop. Hence early diagnosis is of the greatest value.

In individuals who have suffered from syphilis, it is important to recognize the early or premonitory symptoms of spinal syphilis, and at once to commence vigorous treatment.

In all cases anti-syphilitic treatment should be thoroughly carried out. The form which is most commonly prescribed is the combination of mercurial inunctions with potassium iodide internally. A drachm or

more of the blue (mercurial) ointment may be rubbed into the skin daily, until the patient has been brought well under the influence of the drug, and the ointment continued in quantities sufficient to maintain the action, but to avoid the toxic effects. The ointment may be rubbed into the skin of the limbs in succession (one limb each day). It is well to rub the ointment downwards, towards the distal part (i.e. in the direction of the hairs). When the inunctions are made in this direction, irritation of the hair follicles is less likely to occur. The oleate of mercury is the preparation which Sir William Gowers prefers. One drachm of the 10 per cent. oleate should be rubbed into the skin twice a day for three days, and then continued once a day until the end of a week. If the gums are not sore, the inunction should be resumed twice a day until they are affected. The same small piece of flannel should always be employed for making the inunction, and the amount of oleate used for the first inunction should be two drachms. If a fresh piece of flannel be used each time it will retain more than half of the ointment (Gowers).

Mercury may be given in the form of pill, powder, or mixture, if preferred, when it is necessary to bring the patient rapidly under its influence. When the patient is not likely to carefully follow out the inunction treatment, then subcutaneous or intravenous injections of mercurial preparations may be employed.

Hutchinson recommends the grey powder (Hydrargyrum cum cretâ) in small doses for a long period. He advises one grain of this powder in a pill combined with opium ($\frac{1}{5}$ to $\frac{1}{4}$ of a grain), so as to avoid diarrhœa at the onset of the treatment. He prescribes this pill four, five, six, or seven times a day, and thinks that by dividing the doses toxic symptoms are avoided, and the desired effect produced with much greater certainty.

The two important evil effects of mercury are salivation and diarrhœa ; the latter can be avoided by the use of opium, the former by allowing the patient to suck a small piece of alum for a short time every day, or by the use of an alum mouth wash, and by keeping the teeth well cleaned. With care, it is possible to continue a thorough mercurial treatment for a long period without causing any bad effects.

Five or ten grains of the iodide of potassium or sodium may be given three times a day at first, and then the dose may be increased to 10 or 20 grains, or even a much larger quantity. Sir William Gowers usually gives the iodide of potassium in 10 gr. doses, and has obtained no evidence of the value of larger doses "except in patients who have long been taking moderate doses."

Occasionally iodides are badly borne, even 10 grain doses cannot be taken without causing great gastric irritation or other symptoms. It may then be given in milk or seltzer water and milk. I have found that 5 grains (followed by a little milk) every two hours has been easily taken, and thus 30 grains could be given in the twenty-four hours without any bad effects to patients who could not take the drug in the ordinary mixtures.

Iodides are often given with aromatic spirits of ammonia, and Hutchinson attaches great importance to the combination; he believes the good effects of the iodides to be increased by the addition of ammonia.

Often a combination of the three iodides, of potassium, sodium and ammonium, along with aromatic spirits of ammonia, answers well.

Many neurologists, who have paid much attention to the treatment of syphilitic disease of the nervous system, think that it is not advisable to give iodide and mercury *together* in full doses, except for a short time and in very urgent cases, since there is reason to believe that iodide promotes the elimination of mercury. Hence, after the two drugs have been employed together for a time, it is best to use them alternately—mercurial inunction being employed alone, and then discontinued, and iodide given alone.

The affection of the gums is evidence that enough mercury is present in the system. It may then be stopped for a day or two, and continued in smaller quantity, to maintain the effect, for four weeks; then the iodide may be given.

Sir Wm. Gowers regards the long continued treatment by mercury and iodides as a great mistake, and thinks that by the continued use of potassium iodide for months or years the tissues of the patient may become so accustomed to its presence that the drug no longer holds check on the syphilitic processes. The specific treatment should be energetic, brief, and renewed, but *not* continuous. It should be discontinued at the end of eight weeks or so, and renewed after two, four, or six months. The patient should have four weeks' treatment with iodide every four months during the first year after any true specific symptoms, and every six months for the next three or four years (Gowers).

It is important that the patient should have good nourishing food. Exposure to cold, over-strain, and sexual excess should be avoided. Marriage ought to be forbidden, even when the course of the spinal syphilis has been favourable. The use of electricity—galvanic current—is probably of some slight service when legs are paralysed and flaccid. Gentle massage and passive movements of the legs are also helpful.

A warm daily bath is advisable whilst the patient is under mercurial treatment.

The attention to the condition of the bladder and the prevention of bed-sores are two points of the greatest importance, just as in cases of non-syphilitic myelitis.

For the prevention of bed-sores the use of a water-bed is of great service. If there be retention of urine the catheter should be used, but the most strict precautions should be taken to keep it perfectly aseptic. If cystitis should appear the bladder should be washed out with some antiseptic lotion (containing boracic acid or sodium salicylate), and urotropin may be given internally. All that has been said on pp. 137-8, with reference to the prevention and treatment of cystitis and bed-sores in myelitis, applies equally in spinal syphilis.

Erb states that he has found the use of the hypodermic injection of strychnine of great benefit in the treatment of the sequelæ of the disease, especially the spastic paralysis of the legs and the obstinate disorders of the bladder.

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SECTION XIII

TRAUMATIC LESIONS OF THE SPINAL CORD

These affections are strictly surgical, but after the early period they often come under the care of the physician : hence a short summary of their symptoms and pathology is desirable, even in a work devoted to medical diseases of the spinal cord. For a complete account of traumatic spinal lesions the reader is referred to surgical text-books and special works on the subject.

The following are the chief traumatic lesions of the spinal cord :—

1. Fracture and dislocation of the vertebræ, causing compression or contusion of the cord.
2. Wounds of the cord. Punctured and incised wounds, caused by knives, daggers, or pointed instruments. Gunshot wounds.
3. Laceration of the cord.
4. Spinal hæmorrhage and softening ; meningeal hæmorrhage.
5. Concussion of the cord.
6. Traumatic hysteria, traumatic neurosis, " railway spine."
7. Post-traumatic organic diseases.

According to the statistics of Minor, the most frequent forms of traumatic organic cord affections are those associated with lesions of the vertebræ. In the cervical region vertebral dislocation, with cord compression, is the most common form ; in the lumbar region vertebral fracture with compression of the cord.

A cord lesion may be produced by distortion of the vertebræ—by a sudden forced flexion or extension, in which there is incomplete vertebral displacement of momentary duration (diastasis). The articular processes of the upper vertebra (at the seat of the lesion) glide over those of the lower vertebra, and directly return to their normal position. It is stated that the spinal cord may be severely crushed, and even fatal lesions may be caused in this way, without any permanent lesion of the vertebræ being produced.

A splinter of bone from a fractured vertebra sometimes penetrates the spinal meninges and produces lesion of the cord itself.

Fracture of the Vertebræ and Compression of the Spinal Cord.

The body of the vertebra is more frequently fractured than the arch in the dorsal and lumbar regions ; in the cervical region, in about half of the cases the arch is fractured.

The cord is compressed by the displaced bone, but generally the dura mater is not torn through. There is usually some hæmorrhage between the dura mater and the vertebræ; also the vessels of the pia mater may be ruptured and an intra-dural hæmorrhage may occur.

The **symptoms** of compression of the cord, due to fracture of the vertebræ, may be divided into three groups: (1) local, (2) general, (3) spinal.

1. Local. At the seat of the fracture deformity may be seen in the spinous processes posteriorly; but this sign is not always present. It may not occur at first; but may be produced later owing to movements of the patient. Crepitation is seldom obtained, but it may be detected in fracture of a vertebral arch. Pain at the seat of the fracture is often severe; it is increased by pressure, and frequently radiates along the nerve roots situated near the lesion.

2. The general symptoms are those of shock and sometimes there is loss of consciousness at the onset.

3. The spinal symptoms are those of a transverse lesion of the spinal cord at the seat of compression (*see* p. 114). The reflexes are often lost below the lesion. If the fracture be in the cervical or lumbar region, there is often pain radiating into the arms or legs. Persistent erection of the penis (priapism) is a common symptom, especially if the lesion be in the cervical region. The bladder and rectum are usually paralysed. The motor paralysis in the limbs is more marked than the anæsthesia.

In fracture of the first two cervical vertebræ sudden death is frequent; in other cases the patient dies in a few days; in some cases, however, recovery occurs. The symptoms are often indefinite. There is pain in the neck, and the movements of the head are limited and painful. Deformity may be seen in the pharynx or in the neck. Other symptoms are difficulty of swallowing and breathing, and paralysis of the limbs, of the tongue and soft palate. The temperature falls, and the pulse becomes slow.

In fracture of the third and fourth cervical vertebræ death usually occurs at an early period from respiratory paralysis. In fracture between the fourth cervical and the second dorsal vertebræ the symptoms are those of a cervical transverse myelitis.

Below the second dorsal vertebra the symptoms are those of a dorsal or lumbar myelitis, or of a lesion of the cauda equina, according to the region of the fracture.

Dislocation of the Vertebræ and Lesions of the Spinal Cord.

Vertebral dislocation is generally caused by injury; occasionally it is the result of disease (caries, etc.), but in such cases there is often some slight injury.

Most frequently the dislocation is in the cervical region. At the

seat of the dislocation the upper vertebra is usually displaced forwards, and the spine of the lower vertebra is prominent posteriorly.

The dislocation is often caused by a blow on the head or neck, or by a fall in which the neck comes in contact with the edge of some resisting object. Frequently there is a combination of fracture and dislocation.

Symptoms.—In cervical dislocation there is deformity of the vertebral spines posteriorly. The spines deviate to one side in the neck; or the spine just below the dislocation is very prominent; an abnormal prominenee may be seen in the pharynx if the lesion should affect the upper eervieal vertebræ. The head is bent forwards; the chin rests on the sternum; or it approaehes one shoulder. In other cases the head is displaced backwards. The movements of the head are limited and painful.

The spinal symptoms of a transverse lesion of the eord are caused by actual compression, or by spinal eontusion or hæmorrhage or by myelitis.

Sudden death may be caused by rupture of the transverse ligament over the odontoid process, with dislocation of this process and compression of the cord.

Spinal symptoms are occasionally eaused by an incomplete and temporary dislocation, as already mentioned.

Diagnosis of Fracture and Dislocation.—The diagnosis of fracture of the spine is based upon the history of injury and the presence of localised deformity of the spine, with pain. But if spinal deformity is absent, the diagnosis is less definite, since the spinal symptoms may oceur from concussion, hæmorrhage, etc.

When deformity is absent there is often local pain on percussion of the vertebræ, with limitation of movement and radiating pains in the limbs. (Care should be taken not to produce injury to the cord by the examination.)

In dislocation there is also the spinal deformity, with a history of injury. In both fracture and disloeation the spinal cord symptoms are those of a transverse lesion (partial or complete).

In both fracture and dislocation the examination with X rays often affords conclusive evidence of the bone lesion, espeeially in the cervical region. (For further details respeeting spinal fracture and dislocations *see* surgieal text-books.)

Pathological Changes in Compression of the Cord and other Lesions due to Vertebral Fracture or Dislocation.

In slight affections, the cord may present no lesion to the naked eye, except hæmorrhage. If the cord be crushed to a greater extent, it is flattened or constricted at the seat of the vertebral lesion. In the most severe lesions the cord substance is completely, or almost completely, broken through; but the dura mater may appear normal.

On section, in some cases, the outline of the grey substance is changed—the condition of heteropia being seen, as in artificial spinal changes produced post-mortem. (This condition may be distinguished from post-mortem artefacts by the presence of central hæmatomyelia, compound granular cells, and distended blood vessels, and by secondary ascending and descending degeneration.) In other cases the cord sub-

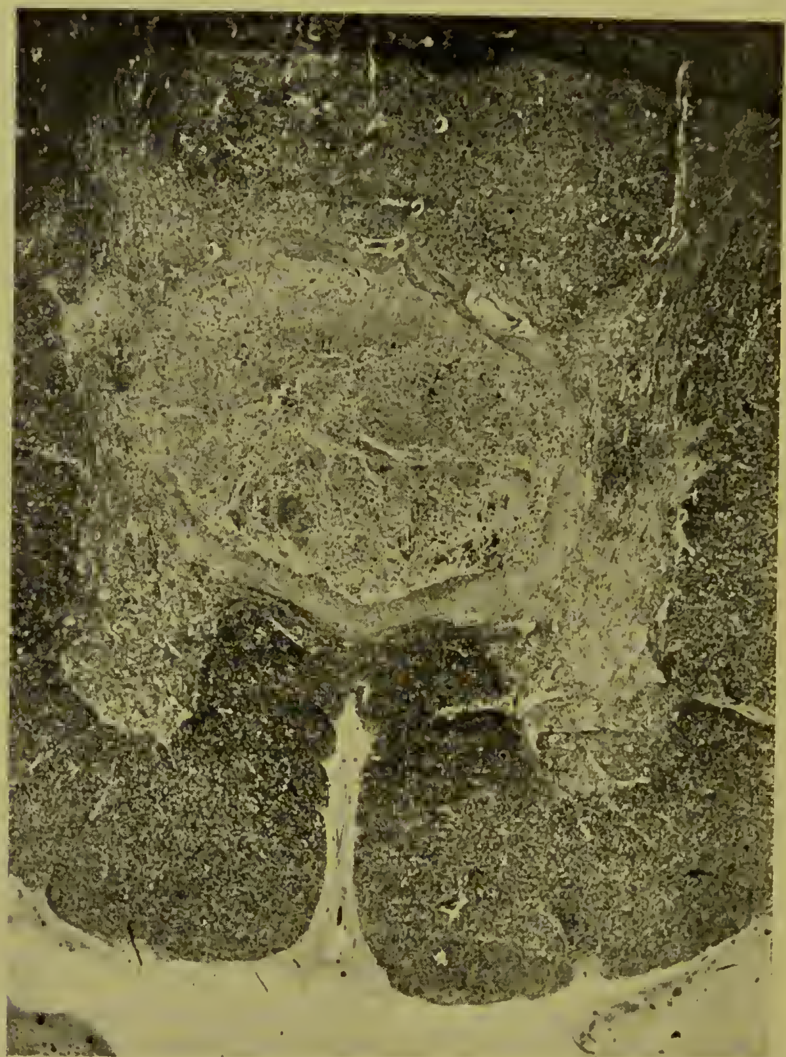

Fig. 183.—Section of Spinal Cord just above Compression Myelitis in a case of fracture-dislocation of vertebræ. Anterior half of Cord seen in section. Weigert's stain. Note greatly dilated central canal (in middle of figure) filled with granular material.

stance is much softened, or there may be the appearances of hæmorrhagic softening.

The microscopical changes vary according to the stage at which the post-mortem examination is made. At an early period there are the changes of hæmorrhagic softening, and the microscope reveals degeneration of the nerve elements and of the neuroglia—myelin globules, infiltration of the part with leucocytes and compound granular cells, blood extravasation in the lymph sheaths of the vessels and free

in the tissues, and cells containing blood pigment, or myelin drops, or red corpuscles (Schmaus and Sacki).

Later secondary "inflammatory" changes are seen. The blood vessels are dilated and full of blood; the lymph sheaths and pericellular spaces are filled with round cells; the surrounding tissues are infiltrated with round cells; and the nerve elements have degenerated. At a still later period the degeneration products are absorbed and a cicatrix is formed.

The central canal of the spinal cord often becomes dilated at the seat of the pathological changes, whilst at other parts of the cord the canal is normal (*see* Fig. 183).

The dura mater is seldom affected; but if a meningeal hæmorrhage has been produced at the onset, at a later period the dura, arachnoid and cord become adherent, and are bound together by a firm ring of fibrous tissue (adhesive meningitis).

In cases of very difficult labour the spinal column of the child is sometimes injured. When traction is made on the feet of the child by the medical man, the spinal column is sometimes ruptured (i.e. the vertebræ are separated) and cord lesions may be produced. Sub-dural or sub-arachnoid hæmorrhage, or hæmatomyelia may occur.

Puncture and Incised Wounds.

These are caused by knives, daggers, or pointed instruments penetrating the spinal canal from behind. The point of the instrument passes between the vertebral arches, or one or more arches are broken through. These puncture wounds occur most frequently in the cervical region.

Usually one-half of the cord or a smaller section thereof is divided, and hence the symptoms are often those of a unilateral lesion (Brown-Séquard's paralysis); but frequently the symptoms of the unilateral paralysis are incomplete. In rare cases the cord has been completely divided and the symptoms have been those of a transverse lesion (*see* p.141).

Not infrequently in puncture wounds the knife does not enter the cord itself, but pushes the membranes into the cord substance, and thus damages its structure.

If foreign bodies, or micro-organisms, enter the wound of the cord the prognosis is much worse. The wound of the cord is at first filled with blood clot, and myelitis is liable to occur around the lesion.

Gunshot Wounds.

The cord may be injured directly by the bullet or by a splinter of bone, and contusion or laceration may be produced. In other cases hæmorrhage occurs in a part adjacent to the spinal cord, and the latter organ is compressed by the blood clot. Meningeal or intra-medullary hæmorrhage may be produced; and sometimes meningitis is caused by the bullet wound.

The cord may also be damaged by momentary subluxation of the spine at the time of the accident.

Laceration.

The spinal cord may be torn across partially or completely in cases of fracture or dislocation of the vertebræ, in bullet wounds, in cases of temporary dislocation of the spine, and possibly in severe concussion.

Complete laceration is rare. The symptoms are those of a transverse lesion of the cord, the knee-jerks are lost, and priapism is common. Death usually occurs in a few hours or days; or at a later period from cystitis or bed-sores. Recovery is impossible in complete laceration.

Spinal Hæmorrhage and Softening.

In some cases the symptoms occurring after an injury are due to hæmatomyelia. In such cases the vertebral column may be unaffected. The hæmorrhage is usually in the grey matter, and often in the lower cervical region (*see* p. 185).

Meningeal hæmorrhage may also be caused by trauma. It may be caused by fracture-dislocation of a vertebra—the edge of the fractured vertebra rupturing a vein (*see* p. 191).

Central softening may also follow an injury.

Concussion of the Spinal Cord.

It has been already mentioned, that after injuries hæmorrhage may occur in the spinal cord, though there is no gross lesion of the spinal columns, and no compression of the cord by displaced bone.

It has been asserted that concussion of the spinal cord may cause severe spinal symptoms—paralysis of limbs, anæsthesia and symptoms of shock; death may occur, and at the autopsy no gross spinal lesion may be found. In other cases, after a few days, recovery occurs. Such cases have been regarded as the result of concussion of the spinal cord without gross lesion, and the symptoms have been attributed to a "molecular change" in the nerve elements. In concussion of the brain post-mortem examination has often failed to reveal any gross lesion, and the cases of supposed spinal concussion have been attributed to a similar nervous "molecular change" in the cord.

In some of the cases of supposed concussion, in which recovery occurs, the condition may be simply traumatic neurosis.

In other cases of supposed concussion of the spinal cord, the autopsy has shown multiple small hæmorrhages or softening, or sub-acute myelitis. Some writers regard all cases of so-called "concussion" of the spinal cord as due to contusion of the cord, and believe that frequently the pathological changes are multiple minute hæmorrhages. But this explanation does not appear to be satisfactory in all cases. Sometimes the hæmorrhages have been too small to account for the symptoms and in other cases hæmorrhages have been absent.

In many cases described as spinal concussion simple anæmic softening has been found, in the form of small multiple foci. Fissures have been occasionally found in the posterior horns of the grey matter of the cord; these may have been caused originally by hæmorrhage.

Also after injury simple parenchymatous degeneration of the fibres of the posterior and lateral columns, or patches of degeneration, have been found post-mortem.

But as already mentioned, in spinal affections following injury, examination of the cord has sometimes revealed no changes. It is possible that the injury may cause death of the nervous elements from shock, owing to " molecular change," and that microscopical examination may not reveal any histological changes. But if the patient lives for a longer period the altered nerve elements break down and degenerative changes are seen. If the effect of the injury to the cord has been less the nerve elements may regain their function, and recovery may occur in course of time.

Some of the rare cases of " spinal concussion " in which the histological examination is negative, may be due to shock conveyed by the cerebro-spinal fluid to the nerve elements, as Schmaus suggests. Such shock might produce loss of function of nerve elements, followed later by necrosis.

Experiments on rabbits have shown that by repeated concussion of the spinal cord tigrolysis and degeneration of the nerve cells and degeneration of nerve fibres may be ultimately produced; and these experiments give some support to the theory of spinal concussion as a rare cause of spinal symptoms after injury (*see* Schmaus).

Traumatic Neurasthenia ; Traumatic Hysteria ; " Railway Spine."

After injuries ill-defined nervous symptoms sometimes occur which are not due to any organic lesion of the spinal cord, although they are often attributed to spinal disease. In a slightly different form, Thorburn gives the following classification of functional neuroses following trauma :—

I. **Acute Effects.**
   (*a*) General nervous depression—" shock."
   (*b*) Acute hysteria.

II. **Chronic after Effects.**
   (*a*) Neurasthenia.
   (*b*) Chronic hysteria.

After railway accidents ill-defined nervous symptoms are sometimes met with which are not associated with any gross lesion of the spinal cord. The patient is stunned at the time of the accident ; often he continues his journey, but he soon feels weak and commences to suffer from pains in the back and legs. After doing his work for a few days he is obliged to discontinue it. A number of subjective symptoms appear—a feeling of general weakness, nervousness, sleeplessness, loss of memory, inability to fix the attention on business, pain and tenderness in the back and symptoms of spinal irritation.

To these subjective symptoms the name of " railway spine " has been given. Spinal pain may continue for years after an accident:

according to Gowers it may be due to traumatic spinal neuralgia. In many of the cases the symptoms just mentioned are more of cerebral than spinal origin, and Thorburn thinks that the term "railway brain" would be more suitable than railway spine

Well marked hysterical symptoms may be present also, and the whole condition has been regarded as due simply to traumatic hysteria or to neurasthenia. Sometimes the symptoms have been attributed simply to sprain of the muscles of the back.

The spinal symptoms may subside after "compensation" for the injury has been obtained : but on the other hand they often persist for a long time afterwards. At present opinions are divided as to whether these cases are all of the nature of traumatic hysteria or neurasthenia, or whether they are due to some real morbid change caused by the accident.

Post-traumatic Organic Spinal Diseases.

Several diseases of the spinal cord (tabes, chronic myelitis, syringomyelia, spinal tumour, disseminated sclerosis, and chronic pachymeningitis) occasionally develop after an injury, and it is thought that trauma may occasionally play some part in the causation of these diseases.

It is known that trauma may cause hæmatomyelia and some neurologists believe that this condition may be occasionally followed by syringomyelia.

General Survey.

The onset of symptoms in traumatic affections of the spinal cord differs according to the nature of the lesion (as described by Sir Wm. Gowers.)

1. In many cases the paralytic symptoms develop immediately. (Fracture, dislocation, wounds, laceration, hæmorrhage, concussion.)

2. In other cases spinal symptoms are slight or absent at first, but in a few days or weeks symptoms of subacute myelitis develop. Pain and sensory symptoms are prominent.

3. In other rare cases, months after the accident, organic spinal disease develops.

Diagnosis of spinal lesions after injury. Rapid improvement of symptoms is in favour of concussion. Spinal bony deformity and the examination with the X rays may afford clear evidence of a fracture or dislocation. Development of paralysis in a few days or weeks after the accident is in favour of subacute myelitis. Paraplegia and sensory symptoms developing directly after the accident may be due to spinal hæmorrhage.

There is often much difficulty in distinguishing between organic disease and neurasthenia. Incontinence of urine and fæces, girdle sensations, ankle-clonus, rectus-clonus, and the extensor type of plantar reflex are all in favour of organic disease.

Treatment.—The treatment of some cases is surgical, and is described in surgical text-books and monographs, to which the reader is referred. Here only a few points with reference to the cases in which operation is justifiable, require consideration.

If examination with the X rays reveals direct compression of the cord which can be removed surgically, operation is desirable. But in most cases no compression can be detected, and then rest and the use of appliances to prevent displacement of bone are indicated. The opinions of surgeons differ greatly as to when operation should be undertaken. In the rare cases in which the arch of a vertebra is fractured, and displaced bone is compressing the cord, operation is indicated. Thorburn recommends operation when there is lesion of the cauda equina, but not in other cases.

Goldscheider draws the following conclusions:—(1) In recent cases operation is not indicated except when, from fracture of the vertebral arch, fragments of bone are penetrating the cord. (2) When the paralysis persists and deformity due to fracture of a vertebral arch is detected, operation is indicated. (3) The most promising cases for operation are those due to fracture of the lower lumbar vertebræ, which would cause lesion of the cauda equina. (4) Laminectomy is not indicated in blood extravasation in the vertebral canal.

In most cases operation is not advisable, and the treatment should then be the same as in myelitis. Rest is important. The actual cautery is of value at the early stage, applied on each side of the spine at the seat of the lesion.

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SECTION XIV

APPENDIX

Notes on Methods of Pathological Examination of the Spinal Cord

Some knowledge of the results of the chief methods of staining microscopical sections of the spinal cord is now necessary, in order to understand many of the illustrations and descriptions in text-books and in medical literature devoted to spinal diseases. Only a few medical men and students have the time for working at the pathology of spinal diseases practically, but the opportunity of occasionally securing a valuable spinal specimen occurs to most men in the course of practice. Unfortunately, in private practice, when such rare opportunity does occur, the specimen is not infrequently spoiled, because it is not preserved in a suitable hardening fluid.

The following account, of methods of pathological examination of the spinal cord, may be useful to practitioners of medicine and senior students, who wish to work *privately* at the pathology of spinal diseases. It furnishes a short account of the *simpler* methods of carrying out the more important staining processes. The methods described may not always be the best, or those most suitable for *class* purposes, but they are the methods which the writer has found convenient in his own pathological work, and they have been used in preparing the sections from which the illustrations in this book have been taken.

In removing the spinal cord at the autopsy, the simplest and quickest method is to place the body face downwards and to remove the cord from the posterior surface,[1] using a *double saw* for division of the vertebral arches. This instrument consists of two semi-circular saws fixed to a handle. (It is figured in Arnold's catalogue of surgical instruments.) By its use the cord can be removed much more rapidly than with an ordinary saw. In removing the cord from the vertebral canal, it should be lifed up by means of forceps, which fix only the dura mater and do not nip the cord itself.

There are many methods of hardening the spinal cord, but three fluids are most useful: Müller's fluid, formol, and 96 per cent. alcohol.

[1] The method of removal of the cord from the front can be carried out rapidly by those who have had much practice in the use of the special instruments required: but it presents some difficulty to medical men who only occasionally undertake post-mortem work.

In most cases it is best to place small thin slices of the cord in 96 per cent. alcohol, for staining with Nissl's stain or some modification; other pieces in formol, 10 per cent. watery solution ; and the rest of the cord in Müller's fluid (bichromate of potash 2·5, sulphate of sodium 1·0, water 100·0). The hardening in 96 per cent. alcohol is completed in 24 to 36 hours ; in formol a few days are required ; in Müller's fluid three months or longer.

After hardening in Müller's fluid or formol thin transverse slices of the cord are washed in water for 24 hours, then placed in absolute alcohol for 24 hours, and afterwards for 24 hours in a celloidin solution consisting of 2 grammes of celloidin, 15cc. absolute alcohol, 15cc. pure ether (sp. gr. ·720).[1] Specimens hardened in 96 per cent. alcohol are placed in absolute alcohol, without washing in water, and then in celloidin. From the celloidin, the pieces of cord tissue are fixed on to small blocks of cork by means of a few drops of the celloidin solution. After exposure of the specimen in the open air for a few minutes it is found to be adherent to the cork. The piece of cork is then floated, specimen downwards, in 80 per cent. alcohol for 24 hours or longer. The cork is fixed in the clamp of a Reichert's microtome and sections cut, the large knife of the microtome being fixed in an oblique manner, so that as much as possible of its cutting edge is used in making the section. The knife and specimen are kept wet with methylated spirit whilst the sections are being cut. Freezing of the specimen is quite unnecessary with this microtome. The sections are lifted from the knife with a small paint brush and placed in water.

I.—General Stains

One of the best and simplest stains for sections of the spinal cord is *aniline blue black.* In sections stained according to this method the axis cylinders of nerve fibres, the nerve cells and their processes, as well as the neuroglia are stained blue black ; whilst the myelin sheaths of nerve fibres are unstained or simply have a very faint blue-black tint. This method enables us to detect swelling of the axis-cylinder, increase or diminution of the size of the nerve fibre, and sclerosis of neuroglia. Sclerosis causes the affected part to appear darker than the normal region under the low power of the microscope. The presence or absence of nerve cells in the anterior horns can be easily noted by this stain. I have found the following method to answer well :—

The sections are placed for many hours in a *weak* watery or alcoholic solution of the stain, which must be of a deep blue-black colour. I have found it best to allow the section to stain in this dilute solution *for 24 hours.* They are then placed in water for several hours, afterwards in 96 per cent. alcohol for a short time, then in absolute alcohol, creosote, and mounted in Canada balsam.

The *method of van Gieson* is a collective stain by which the various

[1] When very thin sections are desired the specimens are placed in a celloidin solution of half the strength already given for 24 hours, and then for 24 hours in a celloidin solution of full strength.

tissues are differently eoloured. It ean be employed for speeimens hardened in aleohol, as well as for those hardened with Müller's fluid.

To 100cc. of a saturated watery solution of picric acid, is added a saturated watery solution of acid fuchsin until the mixture has a brownish red, ruby, or chocolate colour. (Smaller quantities of the fluid can be prepared.) In this solution sections are placed from 2 to 5 minutes. They are then washed in water and placed in 96 per cent. alcohol until they are differentiated ; the white matter then appears yellow or pale yellowish red, the grey matter brownish red, and the pia mater bright red. When the differentiation is completed, the section is cleared in origanum oil, then placed on a slide, dried with filter paper and mounted in Canada balsam. Different tints of staining can be obtained by mixing the picric acid and fuchsin in different proportions.

In seetions stained aeeording to this method, the pia mater and eonneetive tissue around the blood-vessels are stained bright red, the neuroglia brownish red, the medullary sheaths of the nerve fibres yellow, the axis-eylinders and ganglion eells brownish red. Blood in the blood-vessels is stained greenish yellow. (If it is desired to stain the nuelei of the various tissues, the seetions are first plaeed in hæmatoxylin before van Gieson's stain is used.) It is neeessary to allow the seetions to remain in the hæmatoxylin solution 30 to 60 minutes, and afterwards in water for 24 hours. Nuelei are stained bluish red, and the other tissues as already deseribed.

Van Gieson's method is of great serviee for deteeting inerease of neuroglia—selerosis—in the white matter of the spinal eord. It is also useful as an axis-eylinder stain.

II.—Stain for Nuclei and Cell Infiltration

Combined *logwood and eosin* staining is useful in inflammatory eonditions of the eord when mueh eell infiltration is present, as in aeute myelitis, aeute poliomyelitis, aeute forms of spinal syphilis. The nuelei of eells are stained violet, the eell protoplasm and eonneetive tissue elements pink.

The sections are first stained with hæmatoxylin. (A few drops of a concentrated solution of hæmatoxylin crystals in absolute alcohol is added to a 1 per cent. watery solution of alum until the colour is violet. The solution is ready for use after standing for a few days.) Stain in hæmatoxylin solution for two minutes ; wash in water ; then allow to stand in water 24 hours. Place in absolute alcohol ; then in creosote in which a little eosin has been dissolved ; finally in creosote (without eosin) ; mount in Canada balsam.

III.—Nerve Fibre Stains

Peripheral nerves taken direetly from the dead subjeet may be stained in 1 per eent. osmie aeid solution in the usual manner. The myelin sheath is stained deep blaek. This method answers well in cases of peripheral neuritis. A useful eounter stain, for the nuelei of the fibres, is alum earmine or piero-earmine, whieh may be used after the osmie aeid. In the eentral nervous system this method does not answer. Other stains have to be employed.

There are now a number of exeellent stains for medullated nerve fibres. These are of two kinds :—

1. In some of the methods the medullary sheaths of *healthy* nerve fibres are stained deeply, whilst in tracts of marked degeneration, from which nerve fibres have disappeared owing to disease, the staining is very faint (negative stains as regards degeneration). The best methods are those of Weigert and Pal. Others of the same kind are the methods of Heller and Fraenkel.

2. In another method (Marchi's method) only the *degenerated* fibres are stained, whilst normal fibres are not stained. (Positive stain as regards degeneration.)

Staining of the Medullary Sheath of Normal Nerve Fibres.

Weigert's Method (copper-hæmatoxylin). In this method the outer part of the medullary sheath of the nerve fibre is seen to be stained blue black, the inner part of the medullary sheath is unstained, and the axis cylinder is stained faintly light brown or yellow. The neuroglia, nerve cells, blood-vessels and other structures are stained light brown or yellow. Not only are the nerve fibres of the white matter stained, but the network of fine fibres in the grey matter is also stained blue black, and a beautiful appearance is presented. In a markedly degenerated part of the white matter, the medullary sheaths of the nerve fibres are destroyed. Hence the degenerated area is pale yellow owing to the absence of normal black stained fibres. Tracts of *well-marked* degeneration are thus easily recognized. (*See* Plate I.)

In Weigert's method of staining the tissues are hardened in Müller's fluid. The sections are placed for 48 hours in a solution composed of equal parts of water and of a saturated watery solution of acetate of copper. They are then well washed in water and placed for 24 hours in a solution composed of :—

Hæmatoxylin	1 gramme.
Absolute alcohol	10cc.
Saturated solution of lithium carbonate . . .	1cc.
Distilled water	90cc.

The sections are then washed in water, great care being taken to prevent them being injured as they are very brittle. They are afterwards differentiated in the following solution :—

Borax	2
Potassium ferricyanide (red salt)	2·5
Distilled water	200

(A solution half this strength answers better, when time can be allowed for slow differentiation.) In this fluid the sections remain until the grey matter is clear, light brown or yellow, and well differentiated from the black stained white matter. The sections are then well washed [1] in water, placed in absolute alcohol, creosote, and mounted in Canada balsam.

Staining of Degenerated Nerve Fibres.

Pal's Method. The specimens are hardened in Müller's fluid. The sections are stained for 24 to 48 hours in Weigert's hæmatoxylin solution (of the strength just given) to which two or three drops of a concentrated solution of lithium carbonate have been added. The sections are then well washed in tap water, and if not deep black in colour they are allowed to stand in water for several hours or a day. Afterwards they are placed (one or two at a time) in a ¼ per cent. watery

[1] In order to prevent further change in the sections it is most important that they should be very thoroughly washed several times in water.

solution of potassium permanganate for 20 or 30 seconds or longer, until the grey substance is differentiated and appears yellowish brown. They are then washed in water. (If not sufficiently well differentiated they are placed again in the potassium permanganate solution.) The sections are afterwards placed in the following solution :—Pure oxalic acid 1, potassium sulphite 1, distilled water 200. In a few seconds, a minute, or longer, the grey matter of the section becomes very pale, greyish-white, whilst the white matter appears black. The specimens are then washed well in water. If not dark enough they may be left in tap water for some hours or a day or two ; this deepens the colour.

The medullary sheaths of the nerve fibres are stained black by this method, as in Weigert's stain. Tracts of marked degeneration are unstained, and appear white owing to the absence of nerve fibres. (As a counter stain eosin or alum carmine may be used to colour the neuroglia and degenerated tracts red.) Finally, the sections are placed in absolute alcohol, creosote, and mounted in Canada balsam. (The counter staining by eosin may be carried out by dissolving a little eosin in the alcohol or creosote.)

Marchi's Method. This method is of great value in staining *degenerated* nerve fibres, and is most frequently employed for revealing tracts of degeneration after lesions of the nervous system produced by experiments on animals. It is also of great service in the demonstration of tracts of recently degenerated nerve fibres in spinal affections in man.

In *fresh* specimens of nervous tissue, as already mentioned, osmic acid stains the myelin sheath of the nerve fibres deep black. But, if the specimen has been kept in bichromate of potash for 8 days, osmic acid will no longer stain the myelin sheath of normal nerve fibres. Fat, however, is stained black in such specimens.

When a medullated nerve fibre is separated from its cell of origin by a pathological or experimental lesion, the nerve fibre on the peripheral side of the lesion (i.e. the part no longer in connexion with the nerve cell) degenerates, and the medullary sheath breaks up into fat globules (Wallerian degeneration). In Marchi's method these degenerated products stain deep black with osmic acid after the specimen has been hardened in Müller's fluid, though the normal nerve fibres remain unstained. Hence in Marchi's method the degenerated fibres are stained deep black, whilst the normal fibres only show the faint yellow staining due to the bichromate (positive result as regards degeneration). Fig. V, Plate I. In sections stained according to Weigert's and Pal's method we have the opposite results—normal fibres stained black, degenerated tracts unstained (negative result).

It is important to remember, that there is a definite period in the degeneration, when the Marchi's stain gives the best results. After experimental lesions in animals, the degeneration of nerve fibres cannot be detected at once; it is seen best 3 to 6 weeks after the lesion. In course of time the degenerated products become absorbed, neuroglia tissue develops in their place, and Marchi's stain gives then no result. In man, after pathological lesions, the degeneration is slow, and it cannot be stated so definitely when Marchi's stain will or will not reveal degeneration. Marchi's method may be expected to show the degeneration, when death has occurred not less than one week and not more than six months after the lesion. But in disease the degeneration is often

slow, and Marchi's stain may reveal degenerated fibres long after the onset of the affection. If, however, the disease is so chronic that all the degeneration products have been absorbed, then Marchi's method will give no results. In such cases Weigert's stain would probably reveal the degeneration (negative results). Both methods should therefore be employed, since Marchi's method may or may not show the degeneration according to the stage of pathological changes. Marchi's method is most suitable for recent degeneration of nerve fibres. Weigert's method for chronic degeneration. Marchi's method is as follows :—

The cord is hardened in Müller's fluid for 10 to 15 days or longer. Thin transverse slices, 2–5 mm. in thickness, are placed in a mixture of two volumes of Müller's fluid with one volume of a 1 per cent. solution of osmic acid, and kept in the dark for 6 to 8 days or longer (the fluid being changed after the first and third days.) They are then washed for 24 hours in water : transfered to alcohol, 70 per cent., for 4 to 12 hours (changing the fluid two or three times), then to 95 per cent. alcohol for one hour, afterwards to 97 per cent. alcohol for 1 to 2 hours. The specimens are embedded in celloidin, and sections cut. The sections are cleared in carbol xylol (three parts of xylol and one of carbolic acid) or creosote, and mounted in Canada balsam.

It may be here mentioned, that if the osmic acid is too strong sometimes diffuse black dots may be seen scattered over the sections, but they are not limited to any tract or special structure of the cord. It is advisable to place several pieces of the cord in the mixed bichromate and osmic acid solution, and to remove them at different periods so that sections showing different degrees of staining may be obtained.

IV.—Axis-Cylinder Stains

The aniline blue-black method, already described, is an excellent and simple stain for axis-cylinders. But the neuroglia is also coloured faintly or deeply according to the degree of staining.

In van Gieson's method, already described, the axis-cylinders, neuroglia and connective tissue are all stained. The following method is especially useful for demonstrating axis-cylinders :—

Nitrate of Silver Stain for the Axis-Cylinders of Nerve Fibres.

Faierstain (*Neurolog. Centralblatt*, 1901, No. 3) and Bielschowsky (*Neurolog. Centralblatt*, 1902, No. 13) have published methods of staining the axis-cylinders of nerve fibres by means of formol and nitrate of silver solutions. The method of Faierstain is somewhat complicated, that of Bielschowsky is less elaborate. I have found a little difficulty in carrying out Bielschowsky's process, but by the following slight modification of it, I have obtained excellent results :—

The specimens are hardened in Müller's fluid, embedded in celloidin and cut in the usual way. Sections are placed for 5 minutes in a solution made by adding three drops of commercial formalin (which contains 40 per cent. of formaldehyde) to 10cc. of tap water. The sections are then washed and placed in a silver solution prepared as follows :—Three drops of liquor ammoniæ (the dilute liquor of the British Pharmacopœia) are dropped into a test tube. Then a little nitrate of silver solution (10 per cent. in water) is added, drop by drop, until a brownish precipitate is obtained. This is dissolved by adding more liquor ammoniæ, drop

by drop, until the fluid is quite clear. Then tap water is added to the mixture up to 10cc. (If the solution is finally slightly turbid add one or two drops more of the liquor ammoniæ.) In this solution the sections remain for 5 or 10 minutes. (If left for several hours good results can still be obtained.) The sections are then washed very well in two watch-glasses of water and placed back again in the dilute formalin solution first used. They are kept in this solution until they become *greyish-black*. By practice the right amount of staining can be estimated. If the sections after immersion in the nitrate of silver solution are not kept in the dilute formalin solution long enough, the axis-cylinders are too faintly stained; if kept too long in the formalin the sections are too dark and the neuroglia is stained black as well as the axis-cylinders. Usually 1 to 3 minutes is sufficient, but the sections must be watched carefully. To the naked eye the grey matter should appear differentiated from the white if the staining is satisfactory. By a few trials it is easy to recognise the right degree of staining by watching the sections. When this degree of staining has been obtained the sections are washed in water, then placed in the following gold solution :—To 10cc. of water are added two drops of a 1 per cent. watery solution of chloride of gold, a few drops of a saturated solution of borax, and a few drops of a 10 per cent. solution of carbonate of potash. In the solution the sections are left for a few minutes, then placed for a few minutes in a 10 per cent. watery solution of sodium hyposulphite; afterwards the sections are washed in water, dehydrated in alcohol of increasing strength, and cleared in oil of cajuput. They are then placed on a slide, the oil washed away with xylol, and the sections mounted in Canada balsam. A glass needle should be used for transferring the sections to the various fluids. (The glass needle can be made from a piece of glass tubing, by heating it in a gas flame and drawing out the heated part into a fine thread.)

When the staining is successful the axis-cylinders of nerve fibres are stained deep black. The myelin sheath is unstained. The neuroglia is not stained, or stained only very faintly grey. The nerve cells and their processes and the fine network of fibres in the grey matter are beautifully stained deep black. By the nitrate of silver process, Bielschowsky has shown that it is possible to clearly demonstrate the persistence of axis-cylinders in the sclerosed patches in the spinal cord in disseminated sclerosis.

Ramon y Cajal recommends the following method of staining axis-cylinders. (See *Compt. Rend. Soc. Biol.*, December 12, 1903, p. 1565.)

1. Pieces of cord or brain, 3–4 mm. in thickness are placed in a 3 pc. solution of silver nitrate for 2 to 10 days. They are kept at a temperature of 35° to 40° C.

2. Wash in distilled water for 2 minutes.

3. Place in the following solution :—

    Pyrogallic acid, 1 gramme.
    Distilled water, 100cc.
    Formol (commercial), 5–10cc.

The specimens remain in this solution for 24 hours.

4. Wash in distilled water for 2 minutes.

5. Harden in absolute alcohol, or a 90 per cent. alcohol, and then in 95 per. cent alcohol or absolute alcohol.

6. Embed in celloidin.

7. Cut thin sections; mount in Canada balsam.

The finest nerve fibrillæ are stained reddish brown or black, and the rest of the tissues yellow.

V.—Nerve Cell Stains

The aniline blue-black staining, as already described, answers very well for most purposes as a stain for nerve cells ; but if it is desired to study the condition of the chromophile substance (Nissl's granules), the methylene blue method of Nissl should be employed, or some modification thereof. One of the best and simplest modifications is the toluidin-blue stain. The fellowing method, for the exact details of which I am indebted to Dr. Jacobsohn, of Berlin, I have found very useful and easy :—

Toluidin-blue Stain. Small transverse slices of the cord, 2–5mm. in thickness are hardened in 96 per cent. alcohol for 24 to 36 hours. One surface of the specimen is then cut as smoothly as possible and lightly dried with blotting-paper ; this surface is fixed to a smooth piece of cork by means of a very thin layer of syndetikon. The specimen is allowed to remain exposed to the air for a few minutes until it is firmly adherent to the cork. To prevent the specimen becoming too dry its surface is touched now and then with a drop of 96 per cent. alcohol. The cork is then floated (specimen downwards) in 96 per cent. alcohol for 24 hours. Afterwards sections are cut with a microtome. The sections are stained for 3 to 5 minutes in a solution of toluidin-blue 0·25 in distilled water 100. After this staining they are placed in 96 per cent. alcohol until the section is differentiated, i.e., until the white matter becomes very light blue and the grey matter only retains a very pale blue tint. The sections are then placed in absolute alcohol for one minute, afterwards in xylol and mounted in Canada balsam. Instead of fixing the specimen to cork with syndetikon, I have obtained good results by embedding in celloidin and then staining the sections as already described. Paraffin embedding is recommended, however, for this method of staining. Chromatolysis and the various changes in the chromophile substance are seen very well by this simple staining. See Fig. 1, Plate I.

Golgi's method and its modifications have been of great service in revealing the external structure of normal nerve cells and their processes. The cells and their numerous processes appear black and opaque under the microscope in sections prepared according to these methods. These results are obtained by placing the tissue in dilute nitrate of silver or dilute corrosive sublimate solutions for definite periods, after hardening in chromate solution. But for pathological work these methods are often unreliable. Nerve cells and their processes are well stained by Bielschowsky's nitrate of silver method, already described.

VI.—Neuroglia Stain

The best stains for neuroglia are those of Weigert and Mallory, but they are somewhat troublesome to carry out, and excellent staining of the neuroglia can be obtained by van Gieson's method, which has been already described.

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The various methods of staining have each special advantages for the detection of changes in the different tissue elements of the spinal cord. The following methods are to be recommended :—

1. For the detection of recent degeneration in nerve fibres, Marchi's method (positive stain).

2. For the staining of normal nerve fibres and for the detection of

well-marked tracts of chronic degeneration, Weigert's and Pal's methods (negative stain for degeneration).

3. For the staining of axis-cylinders, the nitrate of silver and formol methods.

4. For the detection of changes in the internal structure of nerve cells, the toluidin-blue method.

5. For the staining of the whole nerve cell and its processes, the nitrate of silver and formol method.

6. For the staining of neuroglia, van Gieson's method. (More complicated are the methods of Weigert and Mallory.)

7. As a general (or collective) stain for axis-cylinders, neuroglia and nerve cells, van Gieson's method or aniline blue black.

8. For the detection of inflammatory changes and cell infiltration, logwood and eosin.

9. For the immediate staining of fresh specimens of nerve fibres, osmic acid.

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